

**UNITED STATES  
SECURITIES AND EXCHANGE COMMISSION**

Washington, D.C. 20549

**SCHEDULE TO**

**Tender Offer Statement under Section 14(d)(1) or 13(e)(1)  
of the Securities Exchange Act of 1934**

**GENTIUM S.p.A.**

(Name of Subject Company (Issuer))

**JAZZ PHARMACEUTICALS ITALY S.r.l.  
JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**  
(Names of Filing Persons (Offerors))

**Ordinary Shares, no par value per share  
and  
American Depositary Shares, each representing one Ordinary Share**  
(Title of Class of Securities)

**The CUSIP number for the Ordinary Shares, which are not traded on U.S. markets, is 37250B922.  
The CUSIP number for the related American Depositary Shares is 37250B14.**  
(CUSIP Number of Class of Securities)

**Suzanne Sawochka Hooper, Esq.  
Executive Vice President and General Counsel  
Jazz Pharmaceuticals Public Limited Company  
c/o Jazz Pharmaceuticals, Inc.  
3180 Porter Drive  
Palo Alto, California 94304  
Tel: (650) 496-3777**

*Copy to:*

**Keith A. Flaum, Esq.  
Jane Ross, Esq.  
James R. Griffin, Esq.  
Weil, Gotshal & Manges LLP  
201 Redwood Shores Parkway  
Redwood Shores, CA 94065  
(650) 802-3000**

(Name, address, and telephone number of person authorized to receive notices and communications on behalf of filing persons)

**CALCULATION OF FILING FEE**

<b>Transaction Valuation*</b>	<b>Amount of Filing Fee**</b>
\$1,011,728,625.00	\$130,310.65

\* For purposes of calculating the filing fee pursuant to Rule 0-11(d) only, the Transaction Valuation was calculated on the basis of (i) the aggregate of 17,749,625 Ordinary Shares, no par value per share, which includes (A) 10,984,130 Ordinary Shares represented by 10,984,130 American Depositary Shares outstanding, and (B) 2,194,494 Ordinary Shares not yet outstanding but underlying outstanding equity awards, in each case not owned by the Filing Persons, that may be purchased in the tender offer, and (ii) the tender offer price of \$57.00 per Ordinary Share and per American Depositary Share.

\*\* The amount of the filing fee, calculated in accordance with Rule 0-11 of the Exchange Act, and Fee Rate Advisory #1 for fiscal year 2014, issued August 30, 2013, is \$128.80 per \$1 million (prorated for amounts less than \$1 million) of the aggregate Transaction Valuation. The filing fee is calculated by multiplying the transaction value by 0.0001288.

Check box if any part of the fee is offset as provided by Rule 0-11(a)(2) and identify the filing with which the offsetting fee was previously paid. Identify the previous filing by registration statement number, or the Form or Schedule and the date of its filing.

Amount Previously Paid: None  
Form or Registration Number: N/A

Filing Party: N/A  
Date Filed: N/A

Check the box if the filing relates solely to preliminary communications made before the commencement of a tender offer.

Check the appropriate boxes below to designate any transactions to which the statement relates:

- Third party tender offer subject to Rule 14d-1.
- Issuer tender offer subject to Rule 13e-4.
- Going-private transaction subject to Rule 13e-3.
- Amendment to Schedule 13D under Rule 13d-2.

Check the following box if the filing is a final amendment reporting the results of the tender offer.

If applicable, check the appropriate box(es) below to designate the appropriate rule provision(s) relied upon:

- Rule 13e-4(i) (Cross-Border Issuer Tender Offer)
  - Rule 14d-1(d) (Cross-Border Third-Party Tender Offer)
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This Tender Offer Statement on Schedule TO (this “**Schedule TO**”) relates to the offer by Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* (“**Purchaser**”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“**Parent**” or “**Jazz Pharmaceuticals**”), to purchase all outstanding shares of ordinary stock, no par value per share (the “**Ordinary Shares**”), and all outstanding American Depositary Shares, each representing one Ordinary Share and evidenced by an American Depositary Receipt (“**ADR**”) issued by The Bank of New York, as depositary (the “**ADSs**”) of Gentium S.p.A., a *società per azioni* incorporated in Italy (the “**Company**” or “**Gentium**”), at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer to Purchase dated December 23, 2013 (the “**Offer to Purchase**”) and in the related ADS Letter of Transmittal (the “**ADS Letter of Transmittal**”) and Share Form of Acceptance (the “**Share Form of Acceptance**”) and, together with the ADS Letter of Transmittal and Offer to Purchase, as amended or supplemented from time to time, the “**Offer**”), which are annexed to and filed with this Schedule TO as Exhibits (a)(1)(A), (a)(1)(B) and (a)(1)(G), respectively. As more specifically outlined below, the information set forth in the Offer to Purchase and the related ADS Letter of Transmittal and Share Form of Acceptance are incorporated by reference herein.

**Item 1. Summary Term Sheet.**

This information is set forth in the section of the Offer to Purchase entitled “SUMMARY TERM SHEET” and is incorporated herein by reference.

**Item 2. Subject Company Information.**

This information is set forth in the sections of the Offer to Purchase entitled “THE TENDER OFFER — Certain Information Concerning the Company,” “THE TENDER OFFER — Terms of the Offer” and “THE TENDER OFFER — Price Range of Ordinary Shares and ADSs; Dividends” and is incorporated herein by reference.

**Item 3. Identity and Background of Filing Person.**

This information is set forth in the sections of the Offer to Purchase entitled “THE TENDER OFFER — Certain Information Concerning Parent and Purchaser” and “Schedule I— Directors and Executive Officers of the Purchaser and Parent” and is incorporated herein by reference.

**Item 4. Terms of the Transaction.**

This information is set forth in the sections of the Offer to Purchase entitled “SUMMARY TERM SHEET,” “THE TENDER OFFER — Terms of the Offer,” “THE TENDER OFFER — Withdrawal Rights,” “THE TENDER OFFER — Procedure for Tendering Ordinary Shares and ADSs,” “THE TENDER OFFER — Acceptance for Payment and Payment for Ordinary Shares and ADSs,” “THE TENDER OFFER — Certain Tax Consequences of the Offer” and “THE TENDER OFFER — Possible Effects of the Offer on the Market for Ordinary Shares and ADSs; Nasdaq Listing; Deposit Agreement; Exchange Act Registration and Margin Regulations” and is incorporated herein by reference.

**Item 5. Past Contacts, Transactions, Negotiations and Agreements.**

This information is set forth in the sections of the Offer to Purchase entitled “THE TENDER OFFER — Certain Information Concerning Parent and Purchaser,” “THE TENDER OFFER — Background of the Offer; Past Contacts or Negotiations with the Company” and “THE TENDER OFFER — The Tender Offer Agreement; Other Agreements” and is incorporated herein by reference.

**Item 6. Purposes of the Transaction and Plans or Proposals.**

This information is set forth in the sections of the Offer to Purchase entitled “SUMMARY TERM SHEET,” “THE TENDER OFFER — Possible Effects of the Offer on the Market for Ordinary Shares and ADSs; Nasdaq Listing; Deposit Agreement; Exchange Act Registration and Margin Regulations,” “THE TENDER OFFER — Purpose of the Offer; Plans for the Company” and “THE TENDER OFFER — The Tender Offer Agreement; Other Agreements” and is incorporated herein by reference.

**Item 7. Source and Amount of Funds or Other Consideration.**

This information is set forth in the sections of the Offer to Purchase entitled “SUMMARY TERM SHEET” and “THE TENDER OFFER — Source and Amount of Funds” and is incorporated herein by reference.

**Item 8. Interest in Securities of the Subject Company.**

This information is set forth in the section of the Offer to Purchase entitled “THE TENDER OFFER — Certain Information Concerning Parent and Purchaser” and is incorporated herein by reference.

**Item 9. Persons/Assets, Retained, Employed, Compensated or Used.**

This information is set forth in the section of the Offer to Purchase entitled “THE TENDER OFFER — Fees and Expenses” and is incorporated herein by reference.

**Item 10. Financial Statements.**

The financial statements of Purchaser and Parent and pro forma financial information disclosing the effect of the transaction are not material to the Offer. The information set forth in the section of the Offer to Purchase entitled “SUMMARY TERM SHEET — Is your financial condition relevant to my decision to tender my Ordinary Shares or ADSs in the Offer?” is incorporated herein by reference.

**Item 11. Additional information.**

This information is set forth in the sections of the Offer to Purchase entitled “SUMMARY TERM SHEET,” “THE TENDER OFFER — Possible Effects of the Offer on the Market for Ordinary Shares and ADSs; Nasdaq Listing; Deposit Agreement; Exchange Act Registration and Margin Regulations,” “THE TENDER OFFER — Certain Information Concerning Parent and Purchaser,” “THE TENDER OFFER — The Tender Offer Agreement; Other Agreements” and “THE TENDER OFFER — Certain Legal Matters; Regulatory Approvals” and is incorporated herein by reference. The information set forth in the Offer to Purchase and the related ADS Letter of Transmittal and Share Form of Acceptance are also incorporated herein by reference.

**Item 12. Exhibits.**

<u>Exhibit Number</u>	<u>Description</u>
(a)(1)(A)	Offer to Purchase, dated December 23, 2013.
(a)(1)(B)	Form of ADS Letter of Transmittal.
(a)(1)(C)	Form of Notice of Guaranteed Delivery.
(a)(1)(D)	Form of Letter to Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees.
(a)(1)(E)	Form of Letter to Clients for use by Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees.
(a)(1)(F)	Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9.
(a)(1)(G)	Form of Share Form of Acceptance.
(a)(5)(A)	Summary Advertisement as published in The Wall Street Journal on December 23, 2013.
(a)(5)(B)	Joint Press Release of Jazz Pharmaceuticals and Gentium issued on December 19, 2013 (incorporated by reference to Exhibit 99.1 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 19, 2013).
(a)(5)(C)	Jazz Pharmaceuticals investor presentation first made available on December 19, 2013 (incorporated by reference to Exhibit 99.2 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 19, 2013).
(a)(5)(D)	Transcript from investor/analyst conference call held on December 19, 2013 (incorporated by reference to Exhibit 99.1 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
(a)(5)(E)	Email from Jazz Pharmaceuticals’ Chief Executive Officer to employees, sent on December 19, 2013 (incorporated by reference to Exhibit 99.2 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
(a)(5)(F)	Letter from Jazz Pharmaceuticals’ Chief Executive Officer to Gentium employees, sent on December 19, 2013 (incorporated by reference to Exhibit 99.3 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).

- (a)(5)(G) Media Standby Statement, first used on December 19, 2013 (incorporated by reference to Exhibit 99.4 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
- (a)(5)(H) Gentium Transaction Internal Communications Q&A, first used on December 19, 2013 (incorporated by reference to Exhibit 99.5 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
- (a)(5)(I) Jazz Pharmaceuticals Overview Presentation, first used on December 20, 2013 (incorporated by reference to Exhibit 99.6 from the Schedule TO-C filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
- (b)(1) Commitment Letter, dated as of December 19, 2013, by and between Jazz Pharmaceuticals and Barclays Bank PLC.
- (b)(2) Amendment No. 1, dated as of June 13, 2013, to the Original Credit Agreement and related Guaranty, by and among Jazz Pharmaceuticals, Inc., Jazz Financing I Limited and Purchaser, as borrowers, Jazz Pharmaceuticals, as guarantor, the Lenders thereto and Barclays Bank PLC, as Administrative Agent, Collateral Agent, L/C Issuer and Swing Line Lender (incorporated by reference to Exhibit 10.1 from the Form 8-K filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on June 13, 2013).
- (d)(1) Tender Offer Agreement, dated as of December 19, 2013, by and among Jazz Pharmaceuticals, Gentium and Purchaser (incorporated by reference to Exhibit 2.1 from the Form 8-K/A filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
- (d)(2) Form of Support Agreement (incorporated by reference to Exhibit 99.3 from the Form 8-K/A filed by Jazz Pharmaceuticals with the Securities and Exchange Commission on December 20, 2013).
- (d)(3) Form of Transition, Amendment and Release Agreement.
- (d)(4) Form of Consultancy Agreement.
- (d)(5) Form of Retention and Amendment Agreement.
- (d)(6) Confidentiality and Nondisclosure Agreement, entered into effective September 30, 2013, by and between Jazz Pharmaceuticals and Gentium.

**ITEM 13. INFORMATION REQUIRED BY SCHEDULE 13E-3.**

Not applicable.

**SIGNATURES**

After due inquiry and to the best of my knowledge and belief, I certify that the information set forth in this statement is true, complete and correct.

**JAZZ PHARMACEUTICALS PUBLIC LIMITED  
COMPANY**

By: /s/ Suzanne Sawochka Hooper

Name: Suzanne Sawochka Hooper

Title: Executive Vice President and General Counsel

**JAZZ PHARMACEUTICALS ITALY S.r.l.**

By: /s/ Fintan Keegan

Name: Fintan Keegan

Title: Director

Date: December 23, 2013

## EXHIBIT LIST

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(a)(1)(B)	Form of ADS Letter of Transmittal.
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(a)(1)(D)	Form of Letter to Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees.
(a)(1)(E)	Form of Letter to Clients for use by Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees.
(a)(1)(F)	Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9.
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(d)(3)	Form of Transition, Amendment and Release Agreement.
(d)(4)	Form of Consultancy Agreement.
(d)(5)	Form of Retention and Amendment Agreement.
(d)(6)	Confidentiality and Nondisclosure Agreement, entered into effective September 30, 2013, by and between Jazz Pharmaceuticals and Gentium.

**Offer To Purchase For Cash**  
**All Outstanding Ordinary Shares and American Depositary Shares**  
**(each American Depositary Share representing one Ordinary Share)**  
**of**  
**GENTIUM S.p.A.**  
**at**  
**\$57.00 per ORDINARY SHARE and per AMERICAN DEPOSITARY SHARE**  
**by**  
**JAZZ PHARMACEUTICALS ITALY S.r.l.**  
**a wholly-owned subsidiary of**  
**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

<b>THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.</b>
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Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* (“Purchaser”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“Parent”), is offering to purchase all outstanding shares of ordinary stock, no par value per share (the “Ordinary Shares”) of Gentium S.p.A., a *società per azioni* incorporated in Italy (the “Company”) and all outstanding American Depositary Shares (“ADSs”), each representing one Ordinary Share and evidenced by an American Depositary Receipt (“ADR”), of the Company, at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs) (the “Offer Price”), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in this Offer to Purchase and in the related Share Form of Acceptance (the “Share Form of Acceptance”) and ADS Letter of Transmittal (the “ADS Letter of Transmittal” and, together with the Offer to Purchase and Share Form of Acceptance, each as may be amended or supplemented from time to time, collectively constitute the “Offer”).

The Offer is being made pursuant to a Tender Offer Agreement, dated as of December 19, 2013 (as it may be amended from time to time, the “Tender Offer Agreement”), by and among Parent, Purchaser and the Company. **Under no circumstances will interest be paid on the Offer Price for the Ordinary Shares or ADSs, regardless of any extension of the Offer or any delay in making payment for the Ordinary Shares and ADSs.**

Consummation of the Offer is subject to various conditions set forth in the Tender Offer Agreement, including, but not limited to the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Ordinary Shares and ADSs that, together with any Ordinary Shares and ADSs owned by Parent or Purchaser or any of their subsidiaries, represents at least 66 2/3% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the time that Purchaser first accepts any Ordinary Shares and ADSs for payment pursuant to the Offer (the “Acceptance Time”), plus (B) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time, in all cases without duplication for Ordinary Shares underlying the ADSs, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying ADSs) (the “Permitted Minimum Condition Modification”). Subject to the terms of the Tender Offer Agreement, Purchaser or Parent reserve the absolute right in its sole discretion to waive any of the specified conditions of the Offer in the case of any Ordinary Shares and ADSs tendered other than the condition set forth above. The Offer is also subject to other conditions set forth in this Offer to Purchase. See Section 14 – “Certain Conditions of the Offer.”



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The Company's Board of Directors (the "Company Board") has determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company's shareholders and other stakeholders of the Company. The Company Board has approved and deemed advisable the Tender Offer Agreement and the transactions contemplated thereby and has recommended that the holders of Ordinary Shares and ADSs accept the Offer and tender their Ordinary Shares and ADSs to Purchaser pursuant to the Offer.

A summary of the principal terms of the Offer appears on pages S-i through S-vii. You should read this entire document carefully before deciding whether to tender your Ordinary Shares or ADSs in the Offer.

**THIS TRANSACTION HAS NOT BEEN APPROVED OR DISAPPROVED BY THE UNITED STATES SECURITIES AND EXCHANGE COMMISSION (WHICH WE REFER TO AS THE "SEC") OR ANY STATE SECURITIES COMMISSION, NOR HAS THE SEC OR ANY STATE SECURITIES COMMISSION PASSED UPON THE FAIRNESS OR MERITS OF SUCH TRANSACTION OR UPON THE ACCURACY OR ADEQUACY OF THE INFORMATION CONTAINED IN THIS OFFER TO PURCHASE, THE SHARE FORM OF ACCEPTANCE OR THE ADS LETTER OF TRANSMITTAL. ANY REPRESENTATION TO THE CONTRARY IS UNLAWFUL AND A CRIMINAL OFFENSE.**

**THE OFFER HAS NOT BEEN SUBMITTED FOR CLEARANCE TO CONSOB (THE ITALIAN SECURITIES AND EXCHANGE COMMISSION). ACCORDINGLY, THE OFFER IS NOT BEING MADE AND WILL NOT BE MADE, DIRECTLY OR INDIRECTLY, IN ITALY, AND NO COPIES OF THIS DOCUMENT OR ANY OTHER DOCUMENT RELATING TO THE OFFER HAVE BEEN OR WILL BE DISTRIBUTED IN ITALY, EXCEPT THAT, AS REQUIRED BY LAW, HOLDERS OF RECORD OR THOSE APPEARING ON A SECURITIES POSITION LISTING AS HOLDERS OF ORDINARY SHARES OR ADSs (INCLUDING, PURSUANT TO AN EXEMPTION AVAILABLE UNDER ITALIAN LAW, THE VERY LIMITED NUMBER OF SUCH HOLDERS LOCATED OR RESIDENT IN ITALY) WILL BE SENT COPIES OF THIS DOCUMENT AND OTHER DOCUMENTS RELATING TO THE OFFER. NEITHER THIS DOCUMENT NOR ANY OTHER DOCUMENT RELATING TO THE OFFER MAY BE DISTRIBUTED TO ANY OTHER PERSON LOCATED OR RESIDENT IN ITALY FOR ANY REASON. NEITHER ANY SUCH DOCUMENT NOR ANY INFORMATION CONTAINED HEREIN OR THEREIN CONSTITUTES AN OFFER TO PURCHASE OR A SOLICITATION OF AN OFFER TO SELL OR AN ADVERTISEMENT OF AN OFFER TO PURCHASE ADSs OR ORDINARY SHARES IN ITALY WITHIN THE MEANING OF ARTICLE 1, PARAGRAPH 1, LETTER (V) OF ITALIAN LEGISLATIVE DECREE N. 58 OF FEBRUARY 24, 1998.**

*The Information Agent for the Offer is:*



105 Madison Avenue  
New York, New York 10016  
(212) 929-5500 (Call Collect)

or

(800) 322-2885 (Toll Free)

Email: [tenderoffer@mackenziepartners.com](mailto:tenderoffer@mackenziepartners.com)

December 23, 2013

## IMPORTANT

If you want to tender all or part of your Ordinary Shares or ADSs, you must do one of the following, so as to be completed and received before the Offer expires:

*In the case of Ordinary Shares held in book-entry form*, all of which are dematerialized and recorded in the shareholders' name in the account held by Monte Titoli S.p.A ("Monte Titoli"), complete and sign the Share Form of Acceptance for the Offer, which is enclosed with this document, in accordance with the instructions contained in the Share Form of Acceptance, mail or deliver the Share Form of Acceptance (or an originally signed facsimile thereof) and any other required documents, to The Bank of New York Mellon, as tender agent (the "Tender Agent") and deliver the Ordinary Shares or cause those shares to be delivered to the Monte Titoli account of Société Générale Securities Services S.p.A. the Tender Agent's Italian custodian (the "Tender Agent's Custodian").

*In the case of ADSs held in book-entry form*, all of which are held through facilities of The Depository Trust Company ("DTC") request your broker or other securities intermediary to effect the transaction for you. DTC and its direct and indirect participants will establish their own cutoff dates and times to accept instructions to tender in the Offer, which will be earlier than the Expiration Date stated in this Offer to Purchase. Therefore, you should contact your broker or other securities intermediary to determine the cutoff date and time that is applicable to you.

*In the case of ADSs held in certificated form*, complete and sign the ADS Letter of Transmittal for the Offer, which is enclosed with this document, in accordance with the instructions contained in the ADS Letter of Transmittal (having your signature on the ADS Letter of Transmittal medallion guaranteed if required by the ADS Letter of Transmittal), mail or deliver the ADS Letter of Transmittal (or an originally signed facsimile thereof) and any other required documents, including the certificate for your ADSs, known as an American Depositary Receipt or ADR, to the Tender Agent.

Any holder of ADSs who desires to tender ADSs pursuant to the Offer and whose ADRs representing such ADSs are not immediately available, or who cannot comply in a timely manner with the procedures for tendering ADSs by book-entry transfer, or who cannot deliver all required documents to the Tender Agent by the expiration date of the Offer, may tender such ADSs pursuant to the Offer by following the procedures for guaranteed delivery described in Section 3 – "Procedures for Tendering Ordinary Shares and ADSs."

Questions and requests for assistance should be directed to MacKenzie Partners, Inc., the Information Agent for the Offer, at the address and telephone numbers set forth on the back cover of this Offer to Purchase. Additional copies of this Offer to Purchase, the related Share Form of Acceptance, ADS Letter of Transmittal, Notice of Guaranteed Delivery and other materials related to the Offer may also be obtained at our expense from the Information Agent. Additionally, copies of this Offer to Purchase, the related Share Form of Acceptance, ADS Letter of Transmittal, Notice of Guaranteed Delivery and any other material related to the Offer may be found at [www.sec.gov](http://www.sec.gov). You may also contact your broker or other securities intermediary for assistance.

**This Offer to Purchase, Share Form of Acceptance and ADS Letter of Transmittal contain important information, and you should read both carefully and in their entirety before making a decision with respect to the Offer.**

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## SUMMARY TERM SHEET

The information contained in this summary term sheet is a summary only and is not meant to be a substitute for the more detailed description and information contained in the Offer to Purchase, the Share Form of Acceptance, the ADS Letter of Transmittal and the Notice of Guaranteed Delivery. You are urged to read carefully the Offer to Purchase, the Share Form of Acceptance, the ADS Letter of Transmittal and the Notice of Guaranteed Delivery in their entirety. Parent and Purchaser have included cross-references in this summary term sheet to other sections of the Offer to Purchase where you will find more complete descriptions of the topics mentioned below. The information concerning the Company contained herein and elsewhere in the Offer to Purchase has been provided to Parent and Purchaser by the Company or has been taken from or is based upon publicly available documents or records of the Company on file with the SEC or other public sources at the time of the Offer. Parent and Purchaser have not independently verified the accuracy and completeness of such information. Parent and Purchaser have no knowledge that would indicate that any statements contained herein relating to the Company provided to Parent and Purchaser or taken from or based upon such documents and records filed with the SEC are untrue or incomplete in any material respect.

### Securities Sought

All outstanding shares of ordinary stock, no par value per share (the “Ordinary Shares”) of Gentium S.p.A., a *società per azioni* incorporated in Italy (the “Company”), and all outstanding American Depositary Shares (the “ADSs”), each representing one Ordinary Share and evidenced by an American Depositary Receipt (“ADR”), of the Company. See Section 1 – “Terms of the Offer.”

### Price Offered Per Ordinary Share or ADS (without duplication for Ordinary Shares underlying ADSs)

\$57.00 in cash, without interest thereon and less any required withholding taxes. See Section 1 – “Terms of the Offer.”

### Scheduled Expiration of Offer

12:00 midnight, New York City time, on the evening of January 22, 2014, unless the Offer is otherwise extended. See Section 1 – “Terms of the Offer.”

### Purchaser

Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* and wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland. See Section 1 – “Terms of the Offer.”

### Who is offering to buy my ADSs?

We are Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* (“Purchaser”) formed for the purpose of making this Offer. We are a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“Parent”).

Unless the context indicates otherwise, in this Offer to Purchase, we use the terms “us,” “we” and “our” to refer to Purchaser and, where appropriate, Parent. We use the term “Parent” to refer to Jazz Pharmaceuticals Public Limited Company alone, the term “Purchaser” to refer to Jazz Pharmaceuticals Italy S.r.l. alone and the terms “the Company” or the “Company” to refer to Gentium S.p.A.

See the “Introduction” to this Offer to Purchase and Section 9 – “Certain Information Concerning Parent and Purchaser.”

### What are the classes and amounts of securities sought in the Offer?

We are offering to purchase all of the outstanding Ordinary Shares and ADSs. In this Offer to Purchase we use the term “Offer” to refer to this offer.

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See the “Introduction” to this Offer to Purchase and Section 1 – “Terms of the Offer.”

### **Why are you making the Offer?**

We are making the Offer because we want to acquire control of, and ultimately the entire equity interest in, the Company. Upon completion of the Offer, Parent intends to delist and, to the extent possible, deregister the Ordinary Shares and ADSs.

### **Who can participate in the Offer?**

The Offer is open to all holders and beneficial owners of Ordinary Shares and ADSs. The Offer is not being made in Italy, and the Offer materials will not be distributed in Italy (except that, as required by law, holders of record or those appearing on a securities position listing as holders of Ordinary Shares or ADSs, including the very limited number of such holders located or resident in Italy, will be sent Offer materials).

### **How much are you offering to pay?**

We are offering to pay \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), in cash, without interest thereon and less any required withholding taxes. We refer to this amount as the “Offer Price.”

See the “Introduction” to this Offer to Purchase.

### **Will I have to pay any fees or commissions?**

If you are the registered holder of Ordinary Shares in an account held at Monte Titoli S.p.A. (“Monte Titoli”) or the registered holder of ADSs on the books of The Bank of New York Mellon and you tender your Ordinary Shares or ADSs to us in the Offer, you will not have to pay brokerage fees, or surrender, withdrawal or cancellation fees or similar expenses. If you own your ADSs through a broker or other securities intermediary, and your securities intermediary tenders your ADSs on your behalf, your securities intermediary may charge you a fee for doing so. You should consult your broker or other securities intermediary to determine whether any charges will apply.

See the “Introduction” to this Offer to Purchase and Section 17 – “Fees and Expenses.”

### **Is there an agreement governing the Offer?**

Yes. Parent, Purchaser and the Company have entered into a Tender Offer Agreement, dated as of December 19, 2013 (as it may be amended from time to time, the “Tender Offer Agreement”). The Tender Offer Agreement provides, among other things, for the terms and conditions of the Offer.

See Section 12 – “The Tender Offer Agreement; Other Agreements” and Section 14 – “Certain Conditions of the Offer.”

### **What are the material United States federal income tax consequences of tendering Ordinary Shares or ADSs?**

If you are a U.S. taxpayer, your receipt of cash for Ordinary Shares or ADSs in the Offer will be a taxable transaction for federal income tax purposes under applicable U.S. laws. You will generally recognize gain or loss, if any, in an amount equal to the difference between the amount of cash you receive in the Offer and your adjusted tax basis in the Ordinary Shares and ADSs you sell in the Offer. That gain or loss generally will be capital gain or loss, and will be long-term capital gain or loss if you have held the Ordinary Shares and/or ADSs for more than one year at the time of the exchange of Ordinary Shares or ADSs for cash. If you do not complete

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and sign a Substitute Form W-9 or a Form W-8BEN or other Form W-8, as applicable, you also may be subject to required backup U.S. federal income tax withholding. See Section 5 – “Certain Tax Consequences of the Offer” for a more detailed discussion of the tax treatment of the Offer. **We urge you to consult with your own tax advisor as to the particular tax consequences to you of the Offer.** Your receipt of cash consideration in exchange for your Ordinary Shares or ADSs may also be subject to Italian capital gains tax.

### **Will you have the financial resources to pay for all of the Ordinary Shares and ADSs that you are offering to purchase in the Offer?**

Yes. We expect to have sufficient funds to pay for all Ordinary Shares and ADSs validly tendered and not withdrawn in the Offer. We estimate that the total amount of funds we will need to purchase all of the Ordinary Shares and ADSs in the Offer and to complete the related transactions, including the payment of fees and expenses in connection with the Offer, will be approximately \$1 billion. We expect Parent or one or more of its subsidiaries to provide us with sufficient funds to purchase all Ordinary Shares and ADSs validly tendered in the Offer and any related fees and expenses. Parent expects to finance such cash requirements with a combination of available cash and proceeds from incremental term loans and revolving loan borrowings under its existing senior secured credit facility.

See Section 10 – “Source and Amount of Funds.”

### **Is your financial condition relevant to my decision to tender my Ordinary Shares or ADSs in the Offer?**

We do not think our or Parent’s financial condition is relevant to your decision whether to tender Ordinary Shares or ADSs and accept the Offer because:

- Purchaser was organized solely in connection with the Offer and, prior to the Expiration Date (as defined below), will not carry on any activities other than in connection with the Offer;
- the Offer is being made for all outstanding Ordinary Shares and ADSs solely for cash; and
- the Offer is not subject to any financing or funding condition.

See Section 10 – “Source and Amount of Funds” and Section 12 – “The Tender Offer Agreement; Other Agreements.”

### **Is there a minimum number of Ordinary Shares or ADSs that must be tendered in order for you to purchase any securities?**

Yes. Consummation of the Offer is subject to various conditions set forth in the Tender Offer Agreement, including, but not limited to, the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Ordinary Shares and ADSs that, together with any Ordinary Shares and ADSs owned by Parent or Purchaser or any of their subsidiaries, represents at least 66 <sup>2</sup>/<sub>3</sub>% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the time that Purchaser first accepts any Ordinary Shares and ADSs for payment pursuant to the Offer (the “Acceptance Time”), plus (B) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time, in all cases without duplication for Ordinary Shares underlying the ADSs, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying the ADSs).

### **How long do I have to decide whether to tender my Ordinary Shares and ADSs in the Offer?**

You will have until the Expiration Date to tender your Ordinary Shares and ADSs in the Offer. The term “Expiration Date” means 12:00 midnight, New York City time, on the evening of January 22, 2014, unless we, in accordance with the Tender Offer Agreement, extend the period during which the Offer is open, in which event the term “Expiration Date” means the latest time and date at which the Offer, as so extended, expires. In addition, pursuant to the Tender Offer Agreement, we are obligated to provide a subsequent offering period for the Offer as described below, which will provide you an additional opportunity to tender your Ordinary Shares and ADSs. The Depository Trust Company (“DTC”) and its direct and indirect participants will establish their own cutoff dates and times to accept instructions to tender in the Offer, which will be earlier than the Expiration Date stated in this Offer to Purchase. Therefore, you should contact your broker or other securities intermediary to determine the cutoff date and time that is applicable to you.

If you cannot deliver everything required to make a valid tender of ADSs by the scheduled expiration of the Offer, you may still participate in the Offer by using the guaranteed delivery procedure that is described in Section 3 – “Procedures for Tendering Ordinary Shares and ADSs” prior to the scheduled expiration of the Offer.

See Section 1 – “Terms of the Offer” and Section 3 – “Procedures for Tendering Ordinary Shares and ADSs.”

### **Can the Offer be extended and under what circumstances?**

The Tender Offer Agreement obligates us to extend the Offer for one or more periods, in increments of up to 20 business days (the precise length of which is in Parent’s sole discretion) if all of the conditions to the Offer are not satisfied or waived at the Expiration Date. We are also required to extend the Offer beyond the initial Expiration Date for any period required by any rule, regulation, interpretation or position of the SEC or its staff or applicable law, in each case, applicable to the Offer. Notwithstanding the foregoing, we are not required to extend the Offer beyond April 19, 2014 (the “Termination Date”), unless either Parent or the Company elects to extend this date to September 19, 2014 in the event that certain specified conditions are the only conditions that are not satisfied or waived on April 19, 2014, and such conditions are reasonably likely to be satisfied by such later date if the parties use their reasonable best efforts to do so.

See Section 1 – “Terms of the Offer” and Section 12 – “The Tender Offer Agreement; Other Agreements.”

### **Will there be a subsequent offering period?**

We are obligated to provide a “subsequent offering period” of between three and 20 business days for the Offer. A subsequent offering period will be an additional period of time beginning after we have purchased all Ordinary Shares and ADSs validly tendered during the Offer, during which holders may tender their Ordinary Shares and ADSs and receive the Offer consideration promptly. There would be no condition to our purchase of Ordinary Shares and ADSs validly tendered during the subsequent offering period, and tendering holders would not be allowed to withdraw their Ordinary Shares or ADSs.

See Section 1 – “Terms of the Offer.”

### **How will I be notified if the Offer is extended?**

If we extend the Offer, we will inform The Bank of New York Mellon, which is the tender agent for the Offer (the “Tender Agent”), of any extension and will issue a press release announcing the extension not later than 9:00 a.m., New York City time, on the next business day after the day on which the Offer was scheduled to expire.

See Section 1 – “Terms of the Offer.”

### **What are the most significant conditions to the Offer?**

The Offer is subject to several conditions, including, among other things:

- the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Ordinary Shares and ADSs that, together with any Ordinary Shares and ADSs owned by Parent or Purchaser or any of their subsidiaries, represents at least 66 <sup>2</sup>/<sub>3</sub>% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time, plus (B) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time, in all cases without duplication for Ordinary Shares underlying the ADSs, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying the ADSs).

The Offer is also subject to a number of other conditions set forth in this Offer to Purchase. We expressly reserve the right to waive any such conditions, but we cannot, without the prior written consent of the Company: (a) decrease the Offer Price; (b) change the form of consideration payable in the Offer; (c) reduce the number of Ordinary Shares and ADSs sought to be purchased in the Offer; (d) amend, modify or supplement any of the conditions to the Offer in a manner that is adverse to the holders of Ordinary Shares or ADSs or impose conditions to the Offer that are in addition to the conditions to the Offer as set forth in Section 14 – “Certain Conditions to the Offer”; (e) amend or waive the Minimum Condition (other than a Permitted Minimum Condition Modification); (f) extend the Offer in any manner other than pursuant to certain terms in the Tender Offer Agreement; or (g) amend any terms of the Offer in any manner adverse to the holders of Ordinary Shares or ADSs.

See Section 14 – “Certain Conditions of the Offer.”

### **How do I tender my Ordinary Shares or ADSs?**

*In the case of Ordinary Shares held in book-entry form*, dematerialized and recorded in the shareholders’ name in the account held by Monte Titoli S.p.A., complete and sign the Share Form of Acceptance for the Offer, which is enclosed with this document, in accordance with the instructions contained in the Share Form of Acceptance, mail or deliver the Share Form of Acceptance (or an originally signed facsimile thereof) and any other required documents to *Société Générale*, the Italian custodian for the Tender Agent (the “Tender Agent’s Custodian”) and deliver the Ordinary Shares or cause those Ordinary Shares to be delivered to the Monte Titoli account of Societe Generale Securities Services S.p.A. the Tender Agent’s Italian custodian (the “Custodian”), in each case to be completed and received prior to the Expiration Date. The Company has represented to Purchaser in the Tender Offer Agreement that all Ordinary Shares are held in book-entry form.

*In the case of ADSs held in book-entry form*, all of which are held through facilities of DTC, request your broker or other securities intermediary to effect the transaction for you.

*In the case of ADSs held in certificated form*, complete and sign the ADS Letter of Transmittal for the Offer, which is enclosed with this document, in accordance with the instructions contained in the ADS Letter of Transmittal (having your signature on the ADS Letter of Transmittal medallion guaranteed if required by the ADS Letter of Transmittal), mail or deliver the ADS Letter of Transmittal (or an originally signed facsimile thereof) and any other required documents, including the certificate for your ADSs, known as an American Depositary Receipt or ADR, to the Tender Agent, to be completed and received prior to the Expiration Date. If you cannot deliver the required document to the Tender Agent by the expiration of the Offer, you may still validly tender your ADSs by having a broker or other securities intermediary, which is a member in good



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standing of the Securities Transfer Agents Medallion Program or other eligible institution that would be eligible according to the Offer, guarantee that the missing items will be received by the Tender Agent within three trading days. However, the Tender Agent must receive the missing items within that three trading day period or your ADSs will not have been validly tendered.

See Section 3 – “Procedures for Tendering Ordinary Shares and ADSs.”

### **Until what time may I withdraw previously tendered Ordinary Shares or ADSs?**

You may withdraw your previously tendered Ordinary Shares or ADSs at any time until the Expiration Date. In addition, if we have not accepted your Ordinary Shares or ADSs for payment by March 24, 2014, you may withdraw them at any time after that date until we accept your Ordinary Shares or ADSs for payment. This right to withdraw will not, however, apply to Ordinary Shares or ADSs tendered in the subsequent offering period. DTC and its direct and indirect participants will establish their own cutoff dates and times to accept instructions to withdraw ADSs from the Offer, which will be earlier than the Expiration Date stated in this Offer to Purchase. Therefore, you should contact your broker or other securities intermediary to determine the cutoff date and time that is applicable to you.

See Section 4 – “Withdrawal Rights.”

### **How do I withdraw previously tendered Ordinary Shares or ADSs?**

To withdraw previously tendered Ordinary Shares or ADSs, you must deliver a written notice of withdrawal, or a facsimile of one, with the required information to the Tender Agent while you still have the right to withdraw Ordinary Shares or ADSs. If you tendered Ordinary Shares or ADSs by giving instructions to a broker or other securities intermediary, you must instruct the broker or other securities intermediary to arrange for the withdrawal of your Ordinary Shares or ADSs.

See Section 4 – “Withdrawal Rights.”

### **Has the Offer been approved by the Company’s Board of Directors?**

Yes. The Company’s Board of Directors (the “Company Board”) has determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company’s shareholders and other stakeholders of the Company. The Company Board has approved and deemed advisable the Tender Offer Agreement and the transactions contemplated thereby and has recommended that the holders of Ordinary Shares and ADSs accept the Offer and tender their Ordinary Shares and ADSs to Purchaser pursuant to the Offer.

A more complete description of the reasons for the Company Board’s approval of the Offer is set forth in the Solicitation/Recommendation Statement on Schedule 14D-9, which will be mailed to you in connection with this Offer to Purchase.

### **Will members of the Company Board or the Company’s executive officers holding Ordinary Shares or ADSs tender into the Offer?**

Yes. In connection with the Tender Offer Agreement, the Company’s directors Gigliola Bertoglio, Joyce Bigio, Laura Ferro, Bobby Sandage and Elmar Schnee, as well as Khalid Islam, Chief Executive Officer of the Company, Salvatore Calabrese, Chief Financial Officer of the Company, Giorgio Mosconi, Vice President, Medical Affairs of the Company, Adrian Haigh, Senior Vice President, Commercial Operations and Carin Heringa, Scientific Director and Senior Vice President of the Company have each entered into a support agreement with Parent, pursuant to which each of them is required to tender into the Offer and not withdraw any

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Ordinary Shares or ADSs owned by them as of the date of the Tender Offer Agreement as well as any additional Ordinary Shares or ADSs they may own after such date, and to vote against alternative acquisition transactions. There are approximately 4,016,608 Ordinary Shares and ADSs (including 1,666,608 ADSs issuable upon the exercise of options) subject to the Support Agreements, representing approximately 23% of the fully diluted number of Ordinary Shares and ADSs.

### **If I do not tender my ADSs but the Offer is completed, what will happen to my ADSs?**

Following consummation of the Offer, the number of ADSs that are available for trading and the number of holders of ADSs may be so small that there will no longer be an active public trading market (or, possibly, any public trading market) for the ADSs. In addition, the ADSs may no longer be eligible to be traded on the Nasdaq Global Select Market (“Nasdaq”) or any other securities exchange, and the Company may be able to cease making filings with the SEC or otherwise cease being required to comply with the SEC’s rules relating to publicly held companies.

Following consummation of the Offer, we intend to cause the Company to delist the ADSs from Nasdaq. We also intend to cause the Company to terminate the existing deposit agreement between the Company and The Bank of New York Mellon, as Depositary, dated as of June 15, 2005 (referred to herein as the “Deposit Agreement”), under which the ADSs were issued. If the Deposit Agreement is terminated, and you still hold ADSs, you will be entitled to receive one Ordinary Share in exchange for each ADS held by you upon surrender of your ADSs. However, at any time after the expiration of one year from the date of termination, the Depositary may sell the Ordinary Shares underlying ADSs that have not been surrendered and hold uninvested the proceeds on behalf of holders. There is no public trading market for the Ordinary Shares, and the Company would not be under any obligation to list the Ordinary Shares or otherwise attempt to create such a market following any such delisting and termination.

We also intend, to the extent possible after consummation of the Offer, to cause the Company to deregister the Ordinary Shares and ADSs, which are currently registered under the Securities Exchange Act of 1934, as amended (the “Exchange Act”), so that the Company will no longer be obligated to file any reports with the SEC.

### **What is the market value of my ADS as of a recent date?**

On December 19, 2013, the last full day of trading before we announced of the Offer, the reported closing sales price of the ADSs on Nasdaq was \$55.65 per ADS. We encourage you to obtain a recent market quotation for ADSs before deciding whether to tender your ADSs or Ordinary Shares. The Company’s Ordinary Shares are not traded on any public market.

See Section 6 – “Price Range of Ordinary Shares and ADSs; Dividends.”

### **Will I have appraisal rights in connection with the Offer?**

There are no appraisal rights available in connection with the Offer. If the ADSs were to be delisted from Nasdaq, it is doubtful that any remaining holders of Ordinary Shares would be entitled to exercise withdrawal rights under Italian corporate law. Holders of ADSs, as such, have no withdrawal rights. Withdrawal rights are similar to appraisal rights in that they provide exercising holders with the right to receive cash for their previously listed securities. However, the amount of cash to which a holder would be entitled upon exercise of withdrawal rights in the event of a delisting would be fixed based on a calculation mandated under Italian corporate law, rather than being established by a neutral arbiter based on a number of factors intended to determine fair value, as is the case generally with appraisal rights. Should a withdrawal right apply, the cash amount to which holders exercising withdrawal rights would be entitled in the event of a delisting would be equal

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to the arithmetic average of the closing prices of the ADSs during the six months preceding publication of the call notice convening the Company's shareholders' meeting approving the delisting.

See Section 16 – "Appraisal Rights."

**Who should I call if I have questions about the Offer?**

You may call MacKenzie Partners, Inc., the information agent for the Offer, collect at (212) 929-5500 or toll free at (800) 322-2885. See the back cover of this Offer to Purchase for additional contact information.

## INTRODUCTION

We, Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* (“Purchaser”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“Parent”), are offering to purchase all outstanding shares of ordinary stock, no par value per share (the “Ordinary Shares”), of Gentium S.p.A., a *società per azioni* incorporated in Italy (the “Company”) and all outstanding American Depositary Shares (the “ADSs”), each representing one Ordinary Share and evidenced by an American Depositary Receipt of the Company, at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs) (the “Offer Price”), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in this Offer to Purchase and in the related Share Form of Acceptance (the “Share Form of Acceptance”) and ADS Letter of Transmittal (the “ADS Letter of Transmittal,” and together with the Offer to Purchase and the Share Form of Purchase, each as may be amended or supplemented from time to time, collectively constitute the “Offer”).

The Offer is being made pursuant to a Tender Offer Agreement, dated as of December 19, 2013 (as it may be amended from time to time, the “Tender Offer Agreement”), by and among Parent, Purchaser and the Company. The Tender Offer Agreement is more fully described in Section 12 – “The Tender Offer Agreement; Other Agreements,” which also contains a discussion of the treatment of stock options, ADSs and Ordinary Shares issued or issuable pursuant to the Company’s stock option plans.

Tendering holders whose Ordinary Shares are registered in their own names in an account with Monte Titoli S.p.A. (“Monte Titoli”) or whose ADSs are registered in their own names on the books of The Bank of New York Mellon, as depositary for the ADSs (the “Depositary”) and who tender directly to The Bank of New York Mellon, which is the tender agent for the Offer (the “Tender Agent”) will not be charged brokerage fees or commissions or, except as set forth in Instruction 6 of the ADS Letter of Transmittal, transfer taxes on the purchase of Ordinary Shares or ADSs by us. Holders who hold their ADSs through a broker or other securities intermediary nominee should consult with that securities intermediary as to whether it charges any service fees to tender the ADSs on their behalf. Holders of Ordinary Shares and ADSs who do not complete and sign a Substitute Form W-9 or a Form W-8BEN or other Form W-8, as applicable, may be subject to required backup U.S. federal income tax withholding. Holders of ADSs who tender their ADSs will not have to pay any surrender, withdrawal or cancellation fees otherwise payable pursuant to the Deposit Agreement. We will pay all charges and expenses of the Information Agent and the Tender Agent for the Offer.

Holders that are not U.S. persons may have to comply with certification procedures to establish non-U.S. status in order to avoid information reporting and backup withholding tax requirements.

**The Company’s Board of Directors (the “Company Board”) has determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company’s shareholders and other stakeholders of the Company. The Company Board has approved and deemed advisable the Tender Offer Agreement and the transactions contemplated thereby and has recommended that the holders of Ordinary Shares and ADSs accept the Offer and tender their Ordinary Shares and ADSs to Purchaser pursuant to the Offer.**

A more complete description of the Company Board’s reasons for authorizing and approving the Tender Offer Agreement and the transactions contemplated thereby, including the Offer, will be set forth in the Company’s Solicitation/Recommendation Statement on Schedule 14D-9 under the Securities Exchange Act of 1934, as amended (the “Exchange Act”), which is being furnished to holders of Ordinary Shares and ADSs in connection with the Offer (the “Schedule 14D-9”). Holders of Ordinary Shares and ADSs should carefully read the information set forth in the Schedule 14D-9, including the information set forth under the sub-headings “Background of the Offer” and “Recommendation of the Company’s Board of Directors; Reasons for the Offer.”

The Offer is conditioned upon, among other things, the satisfaction of the Minimum Condition (as described below). The Minimum Condition requires that the number of Ordinary Shares and ADSs that have been validly

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tendered (other than ADSs tendered by guaranteed delivery where actual delivery has not occurred) and not validly withdrawn prior to the expiration of the Offer when added to the number of Ordinary Shares and ADSs then owned by Parent and its subsidiaries, represent at least 66 2/3% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the time that Purchaser first accepts any Ordinary Shares and ADSs for payment pursuant to the Offer (the "Acceptance Time"), plus (B) an additional number of shares up to (but not exceeding) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time, in all cases without duplication for Ordinary Shares underlying the ADSs, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying the ADSs). The Offer also is subject to other conditions set forth in this Offer to Purchase. See Section 14 – "Certain Conditions of the Offer."

This Offer to Purchase and the related Share Form of Acceptance and ADS Letter of Transmittal contain important information that should be read carefully in its entirety before any decision is made with respect to the Offer.

## **THE TENDER OFFER**

### **1. Terms of the Offer.**

Purchaser is offering to purchase all of the outstanding Ordinary Shares and ADSs at the Offer Price. According to the Company, as of December 16, 2013, there were (i) 15,555,131 Ordinary Shares issued and outstanding, of which 10,984,130 Ordinary Shares were represented by an equal number of ADSs, and (ii) 2,194,494 Ordinary Shares subject to outstanding equity awards of the Company.

Upon the terms and subject to the conditions of the Offer (including, if the Offer is extended or amended, the terms and conditions of such extension or amendment), we will accept for payment and promptly pay for all Ordinary Shares and ADSs validly tendered prior to the Expiration Date and not validly withdrawn as permitted under Section 4 – "Withdrawal Rights."

The Offer is conditioned upon, among other things, the satisfaction of the Minimum Condition and the other conditions described in Section 14 – "Certain Conditions of the Offer."

The Tender Offer Agreement obligates us to extend the Offer for one or more periods, in increments of up to 20 business days (the precise length of which is in Parent's sole discretion) if all of the conditions to the Offer are not satisfied or waived at the Expiration Date. We are also required to extend the Offer beyond the initial Expiration Date for any period required by any rule, regulation, interpretation or position of the SEC or its staff or applicable law, in each case, applicable to the Offer. Notwithstanding the foregoing, we are not required to extend the Offer beyond April 19, 2014 (the "Termination Date") unless either Parent or the Company elects to extend this date to September 19, 2014 (the "Extended Termination Date") in the event that certain specified conditions are the only conditions that are not satisfied or waived on April 19, 2014, and such conditions are reasonably likely to be satisfied if the parties use their reasonable best efforts to do so.

We have agreed in the Tender Offer Agreement that, without the prior written consent of the Company, we will not: (a) decrease the Offer Price; (b) change the form of consideration payable in the Offer; (c) reduce the number of Ordinary Shares and ADSs to be purchased in the Offer; (d) amend, modify or supplement any of the conditions to the offer in a manner that is adverse to the holders of Ordinary Shares or ADSs or impose conditions to the Offer that are in addition to the conditions to the offer as set forth in Section 14 – "Conditions of the Offer;" (e) amend or waive the Minimum Condition (other than a Permitted Minimum Condition Modification); (f) extend the Offer in any manner other than pursuant to certain terms in the Tender Offer Agreement; or (g) amend any terms of the Offer in any manner adverse to the holders of Ordinary Shares or ADSs.

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If we extend the Offer, are delayed in our acceptance for payment of or payment (whether before or after our acceptance for payment for Ordinary Shares and ADSs) for Ordinary Shares or ADSs or are unable to accept Ordinary Shares or ADSs for payment pursuant to the Offer for any reason, then, without prejudice to our rights under the Offer, the Tender Agent may retain tendered Ordinary Shares and ADSs on our behalf, and such Ordinary Shares and ADSs may not be withdrawn except to the extent that tendering shareholders are entitled to withdrawal rights as described in Section 4 – “Withdrawal Rights.” However, our ability to delay the payment for Ordinary Shares and ADSs that we have accepted for payment is limited by Rule 14e-1(c) under the Exchange Act, which requires us to pay the consideration offered or return the securities deposited by or on behalf of shareholders promptly after the termination or withdrawal of the Offer.

Subject to the terms of the Tender Offer Agreement and the applicable rules and regulations of the SEC or other applicable laws, we expressly reserve the right to waive any Offer condition at any time and from time to time, to increase the Offer Price or to make any other changes in the terms and conditions of the Offer. Any extension, delay, termination or amendment of the Offer will be followed as promptly as practicable by public announcement thereof, and such announcement in the case of an extension will be made no later than 9:00 a.m., New York City time, on the next business day after the previously scheduled Expiration Date. Without limiting the manner in which we may choose to make any public announcement, we currently intend to make announcements regarding the Offer by issuing a press release and making any appropriate filing with the SEC.

If we make a material change in the terms of the Offer or the information concerning the Offer or if we waive a material condition of the Offer, we will disseminate additional tender offer materials and extend the Offer, in each case, if and to the extent required by Rules 14d-4(d)(1), 14d-6(c) and 14e-1 under the Exchange Act. The minimum period during which the Offer must remain open following material changes in the terms of the Offer or information concerning the Offer, other than a change in price or a change in percentage of securities sought, will depend upon the facts and circumstances, including the relative materiality of the terms or information changes. In the SEC’s view, an offer should remain open for a minimum of five business days from the date the material change is first published, sent or given to holders of Ordinary Shares and ADSs, and with respect to a change in price or a change in the percentage of securities sought, a minimum 10 business day period generally is required to allow for adequate dissemination to holders of Ordinary Shares and ADSs and investor response.

If, on or before the Expiration Date, we increase the consideration being paid for Ordinary Shares and ADSs accepted for payment in the Offer, such increased consideration will be paid to all holders whose Ordinary Shares and ADSs are purchased in the Offer, whether or not such Ordinary Shares and ADSs were tendered before the announcement of the increase in consideration.

We expressly reserve the right, in our sole discretion, subject to the terms and conditions of the Tender Offer Agreement and the applicable rules and regulations of the SEC, not to accept for payment any Ordinary Shares and ADSs if, at the Expiration Date, any of the conditions to the Offer have not been satisfied or upon the occurrence of any of the events set forth in Section 14 – “Certain Conditions of the Offer.” Under certain circumstances, we may terminate the Tender Offer Agreement and the Offer.

The Tender Offer Agreement also requires Purchaser to commence a subsequent offering period. A subsequent offering period will be an additional period of not less than three business days and up to 20 business days beginning on the next business day following the expiration of the Offer in accordance with Rule 14d-11 promulgated under the Exchange Act, during which any remaining holders of Ordinary Shares and ADSs may tender, but not withdraw, their Ordinary Shares and ADSs and receive the Offer Price. In a subsequent offering period, we will immediately accept and promptly pay for all Ordinary Shares and ADSs that are validly tendered during the subsequent offering period. During a subsequent offering period, tendering holders will not have withdrawal rights.

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We will make a public announcement with respect to the subsequent offering period, and any extension thereof, no later than 9:00 a.m., New York City time, on the next business day following the Expiration Date or date of termination of any prior subsequent offering period.

The Company has provided us with its list of holders of Ordinary Shares, and the Depositary has provided us with a list of ADS holders and a security position listings for the purpose of disseminating the Offer to holders of Ordinary Shares and ADSs. This Offer to Purchase and the related Share Form of Acceptance and ADS Letter of Transmittal, as well as the Schedule 14D-9, will be mailed to record holders of Ordinary Shares and ADSs whose names appear on the securityholder list and will be furnished, for subsequent transmittal to beneficial owners of Ordinary Shares and ADSs, to brokers and other securities intermediaries whose names, or the names of whose nominees, appear on the securityholder list or, if applicable, who are listed as participants in a clearing agency's security position listing.

### **2. Acceptance for Payment and Payment for Ordinary Shares and ADSs.**

Subject to the terms of the Offer and the Tender Offer Agreement and the satisfaction or waiver of the conditions to the offer set forth in Section 14 – “Certain Conditions of the Offer,” we will accept for payment and pay for all Ordinary Shares and ADSs validly tendered and not validly withdrawn pursuant to the Offer as promptly as possible and in any event within three business days after the Expiration Date (such time, the “Offer Closing”). During the subsequent offering period in connection with the Offer, we will immediately accept for payment and promptly pay for all additional Ordinary Shares or ADSs as they are tendered during such subsequent offering period, subject to and in compliance with the requirements of Rule 14d-11(e) under the Exchange Act. Subject to compliance with Rule 14e-1(c) under the Exchange Act, we expressly reserve the right to delay payment for Ordinary Shares and ADSs in order to comply in whole or in part with any applicable law. See Section 15 – “Certain Legal Matters; Regulatory Approvals.”

In all cases, we will pay for Ordinary Shares and ADSs accepted for payment pursuant to the Offer only after timely receipt by the Tender Agent of (i) ADRs evidencing such ADSs, confirmation of a book-entry transfer of such ADSs (a “Book-Entry Confirmation”) into the Tender Agent’s account at The Depository Trust Company (“DTC”) or book-entry transfer of such Ordinary Shares to the Monte Titoli account of Société Générale Securities Services S.p.A., the Italian custodian (the “Tender Agent’s Custodian”) for the Tender Agent, pursuant to the procedures set forth in Section 3 – “Procedures for Tendering Ordinary Shares and ADSs,” (ii) in the case of ADSs, the ADS Letter of Transmittal (or a manually signed facsimile thereof), properly completed and duly executed, with any required signature guarantees or, in the case of a book-entry transfer, an Agent’s Message (as defined below) in lieu of the ADS Letter of Transmittal or, in the case of Ordinary Shares, the Share Form of Acceptance (or a manually signed facsimile thereof), properly completed and duly executed, and (iii) any other documents required by the ADS Letter of Transmittal or Share Form of Acceptance.

The term “Agent’s Message” means a message, transmitted by DTC to and received by the Tender Agent and forming a part of a Book-Entry Confirmation, that states that DTC has received an express acknowledgment from the participant in DTC tendering the ADSs that are the subject of such Book-Entry Confirmation, that such participant has received and agrees to be bound by the terms of the ADS Letter of Transmittal and that Purchaser may enforce such agreement against such participant.

For purposes of the Offer, we will be deemed to have accepted for payment, and thereby purchased, Ordinary Shares or ADSs validly tendered and not validly withdrawn as, if and when we give oral or written notice to the Tender Agent of our acceptance for payment of such Ordinary Shares or ADSs pursuant to the Offer. Upon the terms and subject to the conditions of the Offer, payment for Ordinary Shares or ADSs accepted for payment pursuant to the Offer will be made by deposit of the Offer Price for such Ordinary Shares or ADSs with the Tender Agent, which will act as agent for tendering holders for the purpose of receiving payments from us and transmitting such payments to tendering holders whose Ordinary Shares or ADSs have been accepted for payment. If we extend the Offer, are delayed in our acceptance for payment of Ordinary Shares or ADSs or are

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unable to accept Ordinary Shares or ADSs for payment pursuant to the Offer for any reason, then, without prejudice to our rights under the Offer, the Tender Agent may retain tendered Ordinary Shares or ADSs on our behalf, and such Ordinary Shares or ADSs may not be withdrawn except to the extent that tendering shareholders are entitled to withdrawal rights as described in Section 4 – “Withdrawal Rights” and as otherwise required by Rule 14e-1(c) under the Exchange Act. **Under no circumstances will we pay interest on the Offer Price for Ordinary Shares or ADSs, regardless of any extension of the Offer or any delay in making such payment.**

If any tendered Ordinary Shares or ADSs are not accepted for payment pursuant to the terms and conditions of the Offer for any reason, or if ADRs are submitted evidencing more ADSs than are tendered, (i) ADRs representing ADSs which were not purchased will be returned (or ADRs representing ADSs not tendered will be sent), without expense to the tendering shareholder (or, in the case of ADSs tendered by book-entry transfer into the Tender Agent’s account at DTC pursuant to the procedure set forth in Section 3 – “Procedures for Tendering Ordinary Shares and ADSs,” such Ordinary Shares or ADSs will be credited to an account maintained at DTC) and (ii) Ordinary Shares will be returned to the accounts at Monte Titoli from which they were tendered, in each case, as promptly as practicable following the expiration or termination of the Offer.

### **3. Procedures for Tendering Ordinary Shares and ADSs.**

*Valid Tender for Ordinary Shares.* To validly tender Ordinary Shares pursuant to the Offer, a properly completed and duly executed Share Form of Acceptance (or an originally signed facsimile thereof) must be delivered or mailed to the Tender Agent at the address set forth on the Share Form of Acceptance and the Ordinary Shares must be delivered to the Monte Titoli account of the Tender Agent’s Custodian, in each case to be completed and received prior to the Expiration Date.

*Valid Tender for ADSs.* To validly tender ADSs pursuant to the Offer, either (a) a properly completed and duly executed ADS Letter of Transmittal (or an originally signed facsimile thereof) in accordance with the instructions of the ADS Letter of Transmittal, with any required medallion signature guarantees, ADRs evidencing the ADSs to be tendered and any other documents required by the ADS Letter of Transmittal, must be received by the Tender Agent at one of its addresses set forth on the back cover of this document prior to the Expiration Date, (b) such ADSs must be properly delivered pursuant to the procedures for book-entry transfer described below and a confirmation of such delivery received by the Tender Agent (which confirmation must include an Agent’s Message if the tendering holder has not delivered an ADS Letter of Transmittal), prior to the Expiration Date or (c) the tendering holder must comply with the guaranteed delivery procedures set forth below.

*Book-Entry Transfer.* The Tender Agent will establish an account with respect to the ADSs at DTC for purposes of the Offer within two business days after the date of this document. Any financial institution that is a participant in DTC’s systems may make a book-entry transfer of ADSs by causing DTC to transfer such ADSs into the Tender Agent’s account in accordance with DTC’s procedures for such transfer. However, although delivery of ADSs may be effected through book-entry transfer, either the ADS Letter of Transmittal (or an originally signed facsimile thereof), properly completed and duly executed, together with any required medallion signature guarantees, or an Agent’s Message in lieu of the ADS Letter of Transmittal, and any other required documents, must, in any case, be transmitted to and received by the Tender Agent at one of its addresses set forth on the back cover of this document prior to the Expiration Date, or the tendering holder must comply with the guaranteed delivery procedures described below. DTC and its direct and indirect participants will establish their own cutoff dates and times to accept instructions to tender in the Offer, which will be earlier than the Expiration Date stated in this Offer to Purchase. Therefore, you should contact your broker or other securities intermediary to determine the cutoff date and time that is applicable to you. The confirmation of a book-entry transfer of ADSs into the Tender Agent account at DTC as described above is referred to herein as a “Book-Entry Confirmation.” The ADS Letter of Transmittal and any other documents required therein, must be transmitted to and received by the Tender Agent at one of the addresses set forth on the back cover of this document. **Delivery of documents to DTC in accordance with DTC’s procedures does not constitute delivery to the Tender Agent.**



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*Signature Guarantees and Stock Powers.* Except as otherwise provided below, all signatures on a ADS Letter of Transmittal must be guaranteed by a financial institution (including most commercial banks, savings and loan associations and brokerage houses) that is a member in good standing of a recognized medallion program approved by the Securities Transfer Association, Inc., including the Security Transfer Agents Medallion Program, the New York Stock Exchange Medallion Signature Program and the Stock Exchange Medallion Program (each, an “Eligible Institution”). Medallion guarantees from Eligible Institutions are generally not available outside the United States. If you need and are unable to obtain a medallion guarantee from an Eligible Institution, please contact the Information Agent for instructions. See Instruction 5 of the ADS Letter of Transmittal. Signatures on the ADS Letter of Transmittal need not be guaranteed (a) if the ADS Letter of Transmittal is signed by the registered owner of the ADSs tendered therewith and such registered owner has not completed the box entitled “Special Payment Instructions” on the ADS Letter of Transmittal or (b) if such ADSs are tendered for the account of an Eligible Institution. See Instructions 1 and 5 of the ADS Letter of Transmittal. If the ADRs evidencing tendered ADSs are registered in the name of a person other than the signer of the ADS Letter of Transmittal, or if payment is to be made to a person other than the registered owner of the ADRs surrendered, then the tendered ADRs must be endorsed or accompanied by appropriate stock powers, signed exactly as the name or names of the registered owner(s) or holder(s) appear on the certificates, with the signatures on the certificates or stock powers medallion guaranteed as described above.

*Guaranteed Delivery.* A holder who desires to tender ADSs pursuant to the Offer and whose ADRs representing such ADSs are not immediately available, or who cannot deliver the ADRs and all other required documents to the Tender Agent prior to the Expiration Date or who cannot comply with the procedure for book-entry transfer on a timely basis, may tender such ADSs by satisfying all of the requirements set forth below:

- such tender is made by or through an Eligible Institution;
- a properly completed and duly executed Notice of Guaranteed Delivery, substantially in the form made available by us, is received prior to the Expiration Date by the Tender Agent as provided below; and
- ADRs for all tendered ADSs, in proper form for transfer (or a Book-Entry Confirmation with respect to all such ADSs), together with a properly completed and duly executed ADS Letter of Transmittal (or an originally signed facsimile thereof), with any required medallion signature guarantees (or, in the case of a book-entry transfer, an Agent’s Message in lieu of the ADS Letter of Transmittal), and any other required documents, are received by the Tender Agent within three trading days after the date of execution of such Notice of Guaranteed Delivery. A “trading day” is a day on which Nasdaq is open for business.

The Notice of Guaranteed Delivery may be transmitted by facsimile transmission or mailed to the Tender Agent and must include a guarantee by an Eligible Institution in the form set forth in the form of Notice of Guaranteed Delivery made available by Purchaser.

**The method of delivery of ADSs, the ADS Letter of Transmittal and all other required documents, including delivery through DTC, is at the election and risk of the tendering holder. Delivery of all such documents will be deemed made only when such documents are actually received by the Tender Agent (including, in the case of a book-entry transfer, by Book-Entry Confirmation). If such delivery is by mail, it is recommended that all such documents be sent by properly insured registered mail with return receipt requested. In all cases, sufficient time should be allowed to ensure timely delivery.**

*Other Requirements.* Notwithstanding any provision hereof, payment for Ordinary Shares and ADSs accepted for payment pursuant to the Offer will in all cases be made only after timely receipt by the Tender Agent of (a) the ADRs evidencing (or a timely Book-Entry Confirmation with respect to) such ADSs, or a confirmation of delivery of the Ordinary Shares to the Monte Titoli account of the Tender Agent’s Custodian (b) in the case of ADSs, an ADS Letter of Transmittal (or an originally signed facsimile thereof), properly completed and duly executed, with any required medallion signature guarantees (or, in the case of a book-entry

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transfer, an Agent's Message in lieu of the ADS Letter of Transmittal) or, in the case of Ordinary Shares, a Share Form of Acceptance that is properly completed and duly executed and (c) any other documents required by the ADS Letter of Transmittal or Share Form of Acceptance. **Under no circumstances will we pay interest on the Offer Price of the Ordinary Shares or ADSs, regardless of any extension of the Offer or any delay in making such payment.**

*Binding Agreement.* The acceptance for payment by the Purchaser of Ordinary Shares and ADSs tendered pursuant to one of the procedures described above will constitute a binding agreement between the tendering holder and Purchaser upon the terms and subject to the conditions of the Offer.

*Appointment as Proxy.* By executing and delivering a Share Form of Acceptance or ADS Letter of Transmittal as set forth above (or, in the case of a book-entry transfer, by delivery of an Agent's Message, in lieu of an ADS Letter of Transmittal), the tendering holder irrevocably appoints designees of Purchaser as such holder's proxies, each with full power of substitution, to the full extent of such holder's rights with respect to the Ordinary Shares or ADSs tendered by such holder and accepted for payment by Purchaser and with respect to any and all other Ordinary Shares or ADSs or other securities issued or issuable in respect of such Ordinary Shares or ADSs on or after the date hereof. All such proxies and powers of attorney will be considered coupled with an interest in the tendered Ordinary Shares or ADSs. Such appointment is effective when, and only to the extent that, Purchaser accepts for payment Ordinary Shares or ADSs tendered by such holder as provided herein. Upon the effectiveness of such appointment, all prior powers of attorney, proxies and consents given by such holder will be revoked, and, subject to applicable law, no subsequent powers of attorney, proxies and consents may be given (and, if given, will not be deemed effective). The Purchaser's designees will, with respect to the Ordinary Shares, ADSs or other securities and rights for which the appointment is effective, be empowered to exercise all voting and other rights of such holder as they, in their sole discretion, may deem proper in connection with any annual, special, adjourned or postponed meeting of shareholders, by written consent in lieu of any such meeting or otherwise. The Purchaser reserves the right to require that, in order for Ordinary Shares or ADSs to be deemed validly tendered, immediately upon Purchaser's payment for such Ordinary Shares or ADSs, Purchaser must be able to exercise full voting, consent and other rights to the extent permitted under applicable law with respect to such Ordinary Shares or ADSs, including voting at any meeting of shareholders.

*Determination of Validity.* All questions as to the validity, form, eligibility (including time of receipt) and acceptance for payment of any tender of Ordinary Shares or ADSs will be determined by us, in our sole discretion, which determination shall be final and binding on all parties. We reserve the absolute right to reject any and all tenders determined by us not to be in proper form or the acceptance for payment of which may, in the opinion of our counsel, be unlawful. We also reserve the absolute right to waive any defect or irregularity in the tender of any Ordinary Shares or ADSs of any particular shareholder, whether or not similar defects or irregularities are waived in the case of other shareholders. No tender of Ordinary Shares or ADSs will be deemed to have been validly made until all defects and irregularities have been cured or waived to our satisfaction. None of Purchaser, Parent, the Tender Agent, the Information Agent (as defined in Section 17 – "Fees and Expenses") or any other person will be under any duty to give notification of any defects or irregularities in tenders or incur any liability for failure to give any such notification. Our interpretation of the terms and conditions of the Offer (including the ADS Letter of Transmittal and Share Form of Acceptance and the instructions thereto) will be final and binding.

*Information Reporting and Backup Withholding.* Payments made to U.S. shareholders of the Company in the Offer generally will be subject to information reporting and may be subject to backup withholding (currently at a rate of 28%). To avoid backup withholding, U.S. shareholders that do not otherwise establish an exemption should complete and return the Form W-9 included in the ADS Letter of Transmittal, certifying that such shareholder is a U.S. person, the taxpayer identification number provided is correct, and that such shareholder is not subject to backup withholding. Non-U.S. shareholders should submit an Internal Revenue Service ("IRS") Form W-8BEN (or other applicable Form W-8) attesting to such shareholder's exempt foreign status in order to qualify for an exemption from information reporting and backup withholding. Backup withholding is not an

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additional tax. Any amounts withheld under the backup withholding rules will be allowed as a refund from the IRS or a credit against a U.S. shareholder's U.S. federal income tax liability provided the required information is timely furnished to the IRS.

### **4. Withdrawal Rights.**

Except as otherwise provided in this Section 4, tenders of Ordinary Shares and ADSs made pursuant to the Offer are irrevocable.

Ordinary Shares and ADSs tendered pursuant to the Offer may be withdrawn at any time prior to the Expiration Date and, unless theretofore accepted for payment by Purchaser pursuant to the Offer, may also be withdrawn at any time after March 24, 2014. Once Purchaser accepts Ordinary Shares or ADSs for exchange pursuant to the Offer, all tenders not previously withdrawn become irrevocable.

For a withdrawal of Ordinary Shares or ADSs to be effective, a signed notice of withdrawal must be timely received by the Tender Agent at one of its addresses set forth on the back cover of this document. Any notice of withdrawal must specify the name of the person having tendered the Ordinary Shares or ADSs to be withdrawn, the number of Ordinary Shares or ADSs to be withdrawn and the name of the record holder of the Ordinary Shares or ADRs representing the ADSs to be withdrawn, if different from that of the person who tendered such Ordinary Shares or ADSs. The signature(s) on the notice of withdrawal must be medallion guaranteed by an Eligible Institution only if the original ADS Letter of Transmittal required a medallion signature guarantee. If Ordinary Shares or ADSs have been tendered pursuant to the procedures for book-entry transfer as set forth in Section 3 – "Procedures for Tendering Ordinary Shares and ADSs," any notice of withdrawal must specify the name and number of the account at Monte Titoli (for Ordinary shares) or DTC (for ADSs) to be credited with the withdrawn ADSs. If ADRs representing the ADSs to be withdrawn have been delivered or otherwise identified to the Tender Agent, the name of the registered owner and the serial numbers shown on such certificates must also be furnished to the Tender Agent as aforesaid prior to the physical release of such certificates.

Withdrawals of Ordinary Shares and ADSs may not be rescinded. Any Ordinary Shares or ADSs properly withdrawn will thereafter be deemed not to have been validly tendered for purposes of the Offer. However, withdrawn Ordinary Shares and ADSs may be re-tendered by again following one of the procedures described in Section 3 – "Procedures for Tendering Ordinary Shares and ADSs" at any time prior to the Expiration Date.

No withdrawal rights will apply to Ordinary Shares or ADSs tendered during a subsequent offering period and no withdrawal rights apply during the subsequent offering period with respect to Ordinary Shares or ADSs tendered in the Offer and accepted for payment. See Section 1 – "Terms of the Offer."

**We will determine, in our sole discretion, all questions as to the form and validity (including time of receipt) of any notice of withdrawal and our determination will be final and binding. None of Purchaser, Parent, the Tender Agent, MacKenzie Partners, Inc., as information agent (the "Information Agent") or any other person will be under any duty to give notification of any defects or irregularities in any notice of withdrawal or incur any liability for failure to give any such notification.**

### **5. Certain Tax Consequences of the Offer.**

#### *Holders of Ordinary Shares or ADSs.*

The following is a summary of U.S. federal and Italian tax consequences to holders of Ordinary Shares or ADSs upon the tender of Ordinary Shares or ADSs for cash pursuant to the Offer. This summary does not purport to be a comprehensive description of all the tax consequences that may be relevant to a decision to tender Ordinary Shares and ADSs in the Offer. In particular, this summary deals only with beneficial owners of Ordinary Shares or ADSs as capital assets and does not address the tax treatment of beneficial owners who may

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be subject to special tax rules, such as banks, tax-exempt entities, insurance companies or dealers in securities or currencies, or persons that hold Ordinary Shares or ADSs as a position in a “straddle” for tax purposes or as part of a “constructive sale” or a “conversion” transaction or other integrated investment comprised of Ordinary Shares or ADSs and one or more other investments, persons related to the Company or its direct and indirect shareholders, persons who received Ordinary Shares or ADSs as compensation, persons who own or owned 10% or more of the voting power of our shares (directly, indirectly or constructively), persons subject to the 3.8% tax on net investment income, or U.S. holders (defined below) whose functional currency is not the U.S. dollar.

This summary is based upon tax laws of the United States and Italy as in effect on the date hereof and all of which are subject to differing interpretations and/or change, possibly with retroactive effect. Holders of Ordinary Shares or ADSs should consult their own advisors as to the U.S., Italian or other tax consequences of the sale of the Ordinary Shares or ADSs in the Offer, including, in particular, the effect of any state, local or other national tax laws, or U.S. estate tax laws.

For purposes of the summary, a U.S. holder means:

- an individual citizen or resident of the United States;
- a corporation (or entity treated as a corporation for U.S. federal income tax purposes) organized in or under the laws of the United States or its political subdivisions;
- an estate, the income of which is includible in gross income regardless of its source; and
- a trust, if a U.S. court is able to exercise primary jurisdiction over the administration of the trust and one or more U.S. persons have the authority to control all substantial decisions of the trust.

For purposes of the current Income Tax Convention between the United States and Italy (the “Income Tax Convention”) and the United States Internal Revenue Code of 1986 (“Code”), beneficial owners of ADRs evidencing ADSs will be treated as the beneficial owners of the Ordinary Shares represented by those ADSs. Beneficial owners of Ordinary Shares or ADSs who are individual citizens or residents of the United States, corporations organized in or under U.S. law, and U.S. partnerships, estates or trusts (to the extent their income is subject to U.S. tax either directly or in the hands of partners or beneficiaries) generally will be considered to be residents of the United States under the Income Tax Convention. Beneficial owners of Ordinary Shares or ADSs who are considered residents of the United States for purposes of the Income Tax Convention, and are not subject to an anti-treaty shopping provision that applies in limited circumstances, are referred to as “U.S. Treaty holders.” Unless otherwise stated, this summary assumes that a U.S. holder is eligible for the benefits of the Income Tax Convention and is a U.S. Treaty holder.

Special rules apply to U.S. holders that are also residents of Italy, and to U.S. holders who are not U.S. Treaty holders. Such persons should consult their tax advisers as to the tax consequences of the exchange of Ordinary Shares or ADSs for cash pursuant to the Offer. The summary does not discuss the treatment of the exchange of Ordinary Shares or ADSs for cash pursuant to the Offer that are held in connection with a permanent establishment or fixed base of a non-resident beneficial owner in Italy.

Adverse U.S. federal income tax consequences may result to U.S. holders who receive cash pursuant to the Offer if the Ordinary Shares or ADSs are treated as to such holder as stock in a passive foreign investment company (“PFIC”), as defined in Section 1297 of the Code. Such adverse tax consequences include taxation of gain recognized in the Offer at rates applicable to ordinary income, subject to an interest charge to compensate for the deferral of tax liability. The Company has represented in the Tender Offer Agreement that neither the Company nor any of its Subsidiaries (i) has ever been classified as a “passive foreign investment company” as defined in Section 1297 of the Code or (ii) as of the Agreement Date (as defined in the Tender Offer Agreement) expects to be classified as a “passive foreign investment company” as defined in Section 1297 of the Code for the current taxable year or the taxable year that includes the Offer Closing (as defined in the Tender Offer Agreement). The Company has given no assurance that the Company or any of its Subsidiaries would not be

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classified as a PFIC for taxable years which include the Agreement Date through the taxable year that includes the Offer Closing. Nor can there be any assurance the IRS would agree that neither the Company nor any of its Subsidiaries were or will be classified as a PFIC for years prior to or including the Offer Closing. U.S. holders should consult their tax advisor as to the adverse U.S. federal income tax consequences that could result from the exchange of Ordinary Shares or ADSs for cash pursuant to the Offer if it is determined that the Company is or was at any time a PFIC during the U.S. holder's holding period. The remainder of this discussion assumes the Company is not, and will not be for any taxable year prior to and including the Offer Closing, a PFIC.

### **Certain U.S. Federal Income Tax Consequences.**

#### *The Exchange.*

The exchange of Ordinary Shares or ADSs for cash pursuant to the Offer generally will be a taxable transaction for U.S. federal income tax purposes, and a U.S. holder who receives cash for Ordinary Shares or ADSs pursuant to the Offer will recognize gain or loss, if any, equal to the difference between the amount of cash received and such U.S. holder's adjusted tax basis in the Ordinary Shares or ADSs. Such gain or loss will be capital gain or loss, and will be long-term capital gain or loss if such U.S. holder's holding period for the Ordinary Shares or ADSs is more than one year at the time of the exchange of such U.S. holder's Ordinary Shares or ADSs for cash. Long-term capital gains recognized by an individual U.S. holder generally are subject to tax at a lower rate than short-term capital gains or ordinary income. There are limitations on the deductibility of capital losses.

#### *Backup Withholding Tax and Information Reporting*

Payments to U.S. holders made pursuant to the Offer will be subject to information reporting and U.S. federal backup withholding tax unless (i) the U.S. holder furnishes an accurate tax identification number or otherwise complies with applicable U.S. information reporting or certification requirements (typically, by completing an IRS Form W-9) or (ii) the U.S. holder is otherwise exempt from backup withholding. Any amounts so withheld may be credited against the U.S. holder's U.S. federal income tax liability. Holders that are not U.S. persons may have to comply with certification procedures to establish non-U.S. status in order to avoid information reporting and backup withholding tax requirements.

### **Certain Italian Tax Consequences**

#### *Capital Gains and Losses.*

The sale of Ordinary Shares or ADSs pursuant to the Offer is a taxable event for Italian tax purposes.

Capital gains and losses are equal to the difference between the consideration received upon sale and the tax basis in the Ordinary Shares or ADSs sold, which is generally equal to the purchase price, increased by any expenses borne in connection with the purchase, other than interest expense. Capital gains and losses are in principle computed on a LIFO basis.

Capital gains realized by holders not resident in Italy on the disposal of a "qualified" shareholding held not in connection with a permanent establishment or fixed base through which such holders carry on or perform business services in Italy are subject to Italian personal or corporate income tax and are included in the holders' taxable income for an amount equal to 49.72% of the capital gain realized. Losses on the disposal of a "qualified" shareholding can be offset against taxable capital gains for a corresponding amount and, if in excess, can be carried forward for up to four years and be offset against capital gains on "qualified" shareholding realized by such person provided that losses are reported in the tax return in the year of disposal. A "qualified" shareholding is constituted by Ordinary Shares or ADSs and/or rights representing more than 25% of the company's total share capital or more than 20% of its share capital voting at an ordinary shareholders meeting. A disposal of a "qualified" shareholding occurs if, in any 12-month period immediately following the date when a

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shareholding meets one of the thresholds illustrated above, the shareholder engages in disposals of Ordinary Shares or ADSs that, individually or in aggregate, constitute a “qualified” shareholding. The taxable gain realized by a non-resident shareholder who is an individual would be subject to progressive personal income tax rates (currently, the marginal tax rate is equal to 43% and may be increased by local surcharges). The taxable gain realized by a non-resident person other than an individual would be subject to corporate income tax, currently levied at a rate of 27.5%.

Generally, a capital gains tax, levied at a rate of 20%, is imposed on gains realized upon the transfer or sale of “non-qualified” shareholdings whether held within or outside Italy. A “non-qualified” shareholding is constituted by an interest in the Company that does not reach the thresholds described above. Losses on the disposal of a “non-qualified” shareholding can be offset against taxable capital gains for a corresponding amount and, if in excess, can be carried forward for up to four years and be offset against capital gains on “non-qualified” shareholding realized by such person provided that losses are reported in the tax return in the year of disposal. However, under Italian law, an exemption applies to capital gains realized by holders not resident in Italy on the disposal of a “non-qualified” shareholding held not in connection with a permanent establishment or fixed base through which such holders carry on or perform business services in Italy provided that the holder is resident of a State that allows for the exchange of information between the tax authorities as listed by the Italian tax authorities and the holder falls in one of the categories of investors identified by the Italian law and the relevant procedural requirements are satisfied.

The above is subject to any provisions of an income tax convention entered into by the Republic of Italy, if the income tax treaty provisions are more favorable to the holder. Pursuant to the income tax convention, a U.S. Treaty holder will not be subject to Italian tax on any capital gains unless such U.S. holder has a permanent establishment or fixed base in Italy to which Ordinary Shares or ADSs are effectively connected. To this end, U.S. Treaty holders that sell Ordinary Shares or ADSs and claim benefits under the Income Tax Convention may be required to produce appropriate documentation establishing that the conditions set out by the Income Tax Convention have been satisfied.

Other countries have executed income tax conventions with Italy. The majority of income tax conventions entered into by Italy provide that capital gains realized from the disposal of Italian securities are subject to tax only in the country of residence of the seller. Provisions of any applicable income tax conventions and conditions to be satisfied should be assessed on a case by case basis.

### *Registration Tax.*

Contracts relating to the transfer of securities are subject to the registration tax as follows: (i) public deeds and notarized deeds are subject to fixed registration tax at a rate of 168 euro (200 euro as from 1 January 2014); (ii) private deeds are only subject to registration tax in case of use or voluntary registration.

### *Tax on Financial Transactions.*

The tender of the Ordinary Shares or ADS would trigger the Italian tax on financial transactions (“FTT”). The tax on financial transactions generally applies to:

- (a) transfer of the ownership of shares and participating financial instruments which have been issued by companies registered in Italy; and
- (b) transfer of the ownership of securities (such as ADSs) representing equity investments referred to in point a) above, irrespective of the country of residence of the issuer of these securities.

The relevant tax criterion is the place of residence (i.e., the place of the registered office) of the issuer of the shares and the participating financial instruments. So, with reference to the transfer of shares and ADSs, the place of residence of the Depository is not relevant. Instead the country of residence of the Company is relevant.

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The place where the transfers are completed and the residence of the contractual counterparties are not relevant in defining taxed transactions. Accordingly, transfers performed outside the Italian territory through foreign intermediaries are also subject to tax.

The tax is determined proportionally by applying a percentage rate to the value of the counter value of the transaction that varies in relation to the market on which the transaction is carried out. In particular, transfers of ownership mentioned in points (a) and (b) above, not carried out in a recognized regulated market or multilateral trading facility, will be subject to tax at a rate of .22% in 2013 (.20% starting from 2014) while transfers carried out on recognized regulated markets and multilateral trading facilities are subject to tax at the .12% percent rate (.10 % starting from 2014). Nasdaq should qualify as a regulated market for such purposes.

The tax is owed by the persons to whom the ownership of shares, financial instruments and securities representing equity investments (such as ADSs) is transferred, regardless of their country of residence and the place where the contract is concluded.

The burden of the tax on the transfer of the Ordinary Shares or ADSs is imposed on the purchaser which is the beneficiary of the transfer of the ownership. The person liable for the assessment and the payment of the tax are in principle the intermediaries either Italian or non-Italian (i.e., banks, trust companies, investment firms and other intermediaries) involved in the execution of the transactions. Where several intermediaries are involved in the execution of the transactions, the tax must be paid by the intermediary that receives the order to execute the transaction directly from the purchaser or the final counterparty.

Non-Italian intermediaries and other non-resident persons involved in the transaction must comply with the obligations deriving from the application of such tax through their Italian permanent establishment, if any. Non-Italian intermediaries and other non-resident persons having no permanent establishment in Italy can appoint a tax representative in order to comply with all the relevant tax obligations. In cases other than the above, the obligations, including the payment of the tax, must be fulfilled directly by the above-mentioned foreign persons.

### **6. Price Range of Ordinary Shares and ADSs; Dividends.**

There is no public market for Ordinary Shares. Purchaser has been supplied no information with respect to any purchases or sales of Ordinary Shares or the terms thereof during the previous two years. The ADSs are currently traded on the Nasdaq Global Select Market ("Nasdaq"), under the symbol "GENT." The ADSs began trading on Nasdaq on May 16, 2006. Prior to this date, the ADSs were traded on the American Stock Exchange, beginning June 16, 2005 and ending on May 15, 2006, the date the ADSs were de-listed from the American

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Stock Exchange. The following table sets forth, for each of the periods indicated, the high and low closing prices per ADS as reported by Nasdaq.

	<u>High</u>	<u>Low</u>
<b>Fiscal Year Ending December 31, 2013</b>		
First Quarter	\$12.59	\$ 7.74
Second Quarter	8.90	7.37
Third Quarter	27.97	7.74
Fourth Quarter (through December 19, 2013)	59.89	26.99
<b>Fiscal Year Ended December 31, 2012</b>		
First Quarter	\$ 9.20	\$ 5.51
Second Quarter	9.75	8.76
Third Quarter	11.16	9.13
Fourth Quarter	12.35	9.92
<b>Fiscal Year Ended December 31, 2011</b>		
First Quarter	\$12.13	\$ 6.88
Second Quarter	10.38	9.05
Third Quarter	9.99	6.00
Fourth Quarter	6.32	5.65

On December 19, 2013, the last full day of trading in the United States before the public announcement of the execution of the Tender Offer Agreement, the closing price of the ADSs on Nasdaq was U.S. \$55.65 per share. On December 20, 2013, the last full day of trading in the United States before the commencement of the Offer, the closing price of the ADSs on the Nasdaq was U.S. \$57.22 per share. Security holders are urged to obtain current market quotations for the ADSs and to review all information received by them from the Company.

The Company has not paid any dividends since its formation. Pursuant to the Tender Offer Agreement, the Company has agreed not to declare, set aside or pay any dividend on, or make any other distribution in respect of, its capital stock or other securities.

### **7. Possible Effects of the Offer on the Market for Ordinary Shares and ADSs; Nasdaq Listing; Deposit Agreement; Exchange Act Registration and Margin Regulations.**

*Possible Effects of the Offer on the Market for Ordinary Shares and ADSs.* The purchase of Ordinary Shares and ADSs pursuant to the Offer will reduce the number of ADSs that might otherwise trade publicly and could adversely affect the liquidity and market value of the remaining Ordinary Shares and ADSs held by the public. The purchase of Ordinary Shares and ADSs pursuant to the Offer can also be expected to reduce the number of holders of Ordinary Shares and ADSs. As discussed below, we intend to cause the Company to delist the ADSs and to terminate the deposit agreement.

*Nasdaq Listing.* According to the Nasdaq published guidelines, the Company may voluntarily withdraw its ADSs from listing on Nasdaq following approval by the Company Board and by the Company's shareholders at an extraordinary shareholders' meeting. To delist voluntarily, the Company will also have to (i) comply with the SEC's rules issued under Section 12(b) of the Exchange Act, which include the filing of an application with the SEC, following which the SEC will publish a notice and accept comments relating to the proposed delisting, and (ii) obtain any corporate approvals required under Italian corporate law. **We intend to cause the Company to apply for delisting of the ADSs after completion of the Offer.**

If the ADSs are delisted, the market for ADSs would be adversely affected.

*Termination of the Deposit Agreement.* The Depositary is obligated to terminate the deposit agreement at any time at the direction of the Company by mailing notice of termination to the holders of ADSs then



outstanding at least 30 days prior to the termination date fixed in such notice. **Following the completion of the Offer, we intend to cause the Company to instruct the Depositary to terminate the deposit agreement.**

On and after the date of termination, holders of ADSs will be entitled to delivery of Ordinary Shares and any other deposited securities represented by the ADSs surrendered upon (i) surrender of such ADSs at the Corporate Trust Office of the Depositary, (ii) payment of the fee of the Depositary for the surrender of such ADSs and (iii) payment of any applicable taxes or governmental charges. If any ADSs remain outstanding after the date of termination, the Depositary will discontinue the registration of transfers of ADSs, will suspend the distribution of dividends to the holders thereof, and will not give any further notices or perform any further acts under the Deposit Agreement, except (a) collecting dividends and other distributions pertaining to the deposited securities, if any, (b) selling rights, if any, as provided in the deposit agreement and (c) delivering Ordinary Shares, together with any dividends or other distributions received with respect thereto and the net proceeds of the sale of any rights or other property, in exchange for ADSs surrendered to the Depositary (after deduction of the fee of the Depositary for the surrender of ADSs, any expenses for the account of the holder of ADSs and any applicable taxes or government charges). At any time after the expiration of one year from the date of termination, the Depositary may sell any remaining Ordinary Shares underlying the ADSs and will hold uninvested the proceeds of any such sale, as well as any other cash it is holding under the deposit agreement for the pro rata benefit of the holders of ADSs which have not theretofore been surrendered and shall have no liability for interest with respect to such proceeds. The Depositary's only obligation will then be to account for the proceeds of the sale and other cash (after deduction of the fee of the Depositary for the surrender of ADSs, expenses for the account of the holder of ADSs, and any applicable taxes or government charges). After termination, the Company's only obligation under the deposit agreement will be with respect to indemnification and to pay certain amounts to the Depositary. The Depositary has advised Purchaser that its general policy is to sell the Ordinary Shares or any securities received in exchange for such Ordinary Shares following the expiration of the one year period from the termination of a deposit agreement, subject to review of each situation on a case by case basis. Since there is no public market for Ordinary Shares, and we do not intend to take steps to create such a market, the Depositary will be limited in its ability to sell the Ordinary Shares following a termination of the deposit agreement. Given the lack of a trading market for the Ordinary Shares, it is not clear at what price the Ordinary Shares would be sold. We may purchase Ordinary Shares from the Depositary in such circumstances.

If the deposit agreement is terminated, holders of ADSs may (i) surrender the ADRs evidencing ADSs to the Depositary in exchange for the Ordinary Shares underlying such ADSs or (ii) not take any action, in which case the Depositary would likely sell the Ordinary Shares after one year as described above.

*Exchange Act Registration.* The Ordinary Shares and ADSs are currently registered under the Exchange Act. The purchase of the Ordinary Shares and ADSs pursuant to the Offer may result in the Ordinary Shares and ADSs becoming eligible for deregistration under the Exchange Act. Registration of the Ordinary Shares and ADSs may be terminated by the Company upon application to the SEC if the outstanding Ordinary Shares and ADSs are not listed on a "national securities exchange" and there are fewer than 300 holders of record of Ordinary Shares and ADSs.

Termination of registration of the Ordinary Shares and ADSs under the Exchange Act would reduce the information required to be furnished by the Company to holders of its securities and to the SEC and would make certain provisions of the Exchange Act, such as the requirement of furnishing annual reports to security holders, no longer applicable with respect to the Company. Furthermore, the ability of "affiliates" of the Company and persons holding "restricted securities" of the Company to dispose of such securities pursuant to Rule 144 under the Securities Act of 1933, as amended, may be impaired or eliminated. If registration of the Ordinary Shares and ADSs under the Exchange Act were terminated, the Ordinary Shares and ADSs would no longer be eligible for continued inclusion on the Federal Reserve Board's list of "margin securities" or eligible for stock exchange listing or reporting on Nasdaq.

**We intend to cause the Company to apply for termination of the registration of the Ordinary Shares and ADSs after consummation of the Offer if the requirements for termination of registration are met.**

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*Margin Regulations.* The Ordinary Shares and ADSs are currently “margin securities” under the regulations of the Board of Governors of the Federal Reserve System, which classification has the effect, among other things, of allowing brokers to extend credit using such Ordinary Shares and ADSs as collateral. Depending upon factors similar to those described above regarding market quotations, the Ordinary Shares and ADSs might no longer constitute “margin securities” for the purposes of the margin regulations, in which event the Ordinary Shares and ADSs would be ineligible as collateral for margin loans made by brokers.

### **8. Certain Information Concerning the Company.**

The summary information set forth below is qualified in its entirety by reference to the Company’s public filings with the SEC (which may be obtained and inspected as described below under “Available Information”) and should be considered in conjunction with the financial and other information in such reports and other publicly available information. Neither Parent nor Purchaser has any knowledge that would indicate that any statements contained in this Offer to Purchase based on such documents and records are untrue. However, neither Parent nor Purchaser assumes any responsibility for the accuracy or completeness of the information concerning the Company, whether furnished by the Company or contained in such filings, or for any failure by the Company to disclose events that may have occurred or that may affect the significance or accuracy of any such information but which are unknown to Parent or Purchaser.

*General.* The Company operates as a *società per azioni* organized under laws of Italy. The address and telephone number of the Company’s registered office and principal place of business is Piazza XX Settembre 2, 22079 Villa Guardia (Como), Italy, telephone number +39-03-1-537-3200. The Company’s agent for service of process in the United States is CT Corporation System, 111 8th Avenue, New York, New York 10011 and the agent’s telephone number is (212) 894-8940. The Company is a biopharmaceutical company focused on the development and manufacture of the Company’s primary product candidate, defibrotide, an investigational drug based on a mixture of single- and double-stranded DNA extracted from pig intestines.

*Available Information.* The Company is subject to the information and reporting requirements of the Exchange Act applicable to a foreign private issuer and, in accordance therewith, is obligated to file reports and other information with the SEC relating to its business, financial condition, information as of particular dates concerning the Company’s directors and officers, information as of particular dates concerning the principal holders of the Company’s securities and any material interests of such persons in transactions with the Company. Such reports and other information are available for inspection at the SEC’s Public Reference Room at 100 F Street, N.E., Washington, D.C. 20549-0213. Please call the SEC at 1-800-SEC-0330 for further information on the public reference room. Copies of such information should be obtained by mail, upon payment of the SEC’s customary charges, by writing to the SEC at 100 F Street, N.E., Washington, D.C. 20549-0213. The SEC also maintains a website at <http://www.sec.gov> that contains reports, proxy statements and other information regarding registrants that file electronically with the SEC.

### **9. Certain Information Concerning Parent and Purchaser.**

Purchaser is an Italian *società a responsabilità limitata* and to date has engaged in no activities, and will engage in no activities, other than those incident to its formation and the Offer. Purchaser is a wholly-owned subsidiary of Parent. The registered office of Purchaser is at via Manzoni 30, Milano, Italy.

The summary information set forth below is qualified in its entirety by reference to Parent’s public filings with the SEC (which may be obtained and inspected as described below under “Available Information”) and should be considered in conjunction with the more comprehensive financial and other information in such reports and other publicly available information. Parent is a public limited company formed under the laws of Ireland. Parent’s principal executive offices are located at Fourth Floor, Connaught House, One Burlington Road, Dublin 4, Ireland. The telephone number of Parent’s principal executive offices is +011-353-1-634-7800.

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Parent is a specialty biopharmaceutical company focused on identifying, developing and commercializing innovative products that address unmet medical needs. On January 18, 2012, the businesses of Jazz Pharmaceuticals, Inc., Parent's predecessor company and Azur Pharma Public Limited Company ("Azur Pharma"), were combined in a merger transaction (the "Azur Merger"), which was accounted for as a reverse acquisition under the acquisition method of accounting for business combinations, with Jazz Pharmaceuticals, Inc. treated as the acquiring company for accounting purposes. As part of the Azur Merger, Azur Pharma changed its name to Jazz Pharmaceuticals plc, a wholly-owned subsidiary of Azur Pharma and merged with and into Jazz Pharmaceuticals, Inc., with Jazz Pharmaceuticals, Inc. surviving the Azur Merger as Parent's wholly-owned subsidiary, and all outstanding shares of Jazz Pharmaceuticals, Inc.'s common stock were canceled and converted into the right to receive, on a one-for-one basis, Parent's ordinary shares. As a result of the Azur Merger, Parent transitioned from being a standalone public Delaware corporation to being a public limited company organized in, and a tax resident of, Ireland.

The name, business address, current principal occupation or employment, five-year material employment history and citizenship of each director and executive officer of Purchaser and Parent and certain other information are set forth on Schedule I hereto.

During the last five years, none of Parent or Purchaser, or, to the best knowledge of Parent and Purchaser, any of the persons listed in Schedule I to this Offer to Purchase, except as set forth therein, (i) has been convicted in a criminal proceeding (excluding traffic violations or similar misdemeanors) or (ii) was a party to any judicial or administrative proceeding (except for matters that were dismissed without sanction or settlement) that resulted in a judgment, decree or final order enjoining the person from future violations of, or prohibiting activities subject to, federal or state securities laws, or a finding of any violation of such laws.

Except as set forth elsewhere in this Offer to Purchase or Schedule I to this Offer to Purchase: (i) none of Parent and Purchaser and, to the knowledge of Parent and Purchaser, the persons listed in Schedule I hereto or any associate or majority-owned subsidiary of Parent, Purchaser or of any of the persons so listed, beneficially owns or has a right to acquire any Ordinary Shares or ADSs or any other equity securities of the Company; (ii) none of Parent, Purchaser and, to the knowledge of Parent and Purchaser, the persons or entities referred to in clause (i) above has effected any transaction with respect to the Ordinary Shares or ADSs or any other equity securities of the Company during the past 60 days; (iii) none of Parent, Purchaser and, to the knowledge of Parent and Purchaser, the persons listed in Schedule I to this Offer to Purchase, has any contract, arrangement, understanding or relationship with any other person with respect to any securities of the Company (including, but not limited to, any contract, arrangement, understanding or relationship concerning the transfer or the voting of any such securities, joint ventures, loan or option arrangements, puts or calls, guaranties of loans, guaranties against loss or the giving or withholding of proxies, consents or authorizations); (iv) during the two years before the date of this Offer to Purchase, there have been no transactions between Parent, Purchaser, their subsidiaries or, to knowledge of Parent and Purchaser, any of the persons listed in Schedule I to this Offer to Purchase, on the one hand, and the Company or any of its executive officers, directors or affiliates, on the other hand, that would require reporting under SEC rules and regulations; and (v) during the two years before the date of this Offer to Purchase, there have been no contracts, negotiations or transactions between Parent, Purchaser, their subsidiaries or, to Parent's and Purchaser's knowledge, any of the persons listed in Schedule I to this Offer to Purchase, on the one hand, and the Company or any of its subsidiaries or affiliates, on the other hand, concerning a merger, consolidation or acquisition, a tender offer or other acquisition of securities, an election of directors or a sale or other transfer of a material amount of assets.

*Additional Information.* Parent is subject to the information and reporting requirements of the Exchange Act and in accordance therewith is obligated to file reports and other information with the SEC relating to its business, financial condition, information as of particular dates concerning Parent's directors and officers, information as of particular dates concerning the principal holders of Parent's securities and any material interests of such persons in transactions with Parent. Such reports, proxy statements and other information are

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available for inspection and copying at the offices of the SEC in the same manner as set forth with respect to the Company in Section 8 of this Offer to Purchase entitled “Certain Information Concerning the Company.”

### **10. Source and Amount of Funds.**

Purchaser estimates that the total amount of funds it will need to purchase all of the Ordinary Shares and ADSs in the Offer and to complete the related transactions, including the payment of fees and expenses in connection with the Offer, will be approximately \$1 billion. Parent (through one of more of its subsidiaries) is expected to provide Purchaser with sufficient funds to purchase all Ordinary Shares and ADSs validly tendered in the Offer and any related fees and expenses. Parent expects to finance such cash requirements with a combination of available cash and proceeds from incremental term loans and revolving loan borrowings under its existing senior secured credit facility.

#### ***Commitment Letter***

On December 19, 2013, Parent entered into commitment letter with Barclays Bank PLC (“Barclays”), pursuant to which, subject to the terms and conditions set forth therein, in connection with the Offer, Barclays has committed to provide a \$500.0 million incremental term facility (the “Incremental Term Loans”). Under the commitment letter, Barclays has the right to syndicate all or a portion of its commitment to one or more financial institutions or other lenders. Any such syndication will be arranged by Barclays, as sole lead arranger, sole bookrunner and sole syndication agent.

A copy of the credit agreement governing Parent’s existing senior secured credit facility, included as Exhibit A to Amendment No. 1 to the existing senior credit facility, and the commitment letter have been filed as Exhibits (b)(2) and (b)(1) to the Schedule TO. Reference is made to such exhibits for a more complete description of the terms and conditions of the existing senior secured credit facility and the commitment letter, and such exhibits are incorporated herein by reference.

#### ***Description of Parent’s Existing Senior Secured Credit Facility***

The existing senior secured credit facility is governed by that certain Credit Agreement, dated as of June 12, 2012 and amended as of June 13, 2013 (the “Credit Agreement”), by and among Parent, as guarantor, three of its wholly-owned subsidiaries, Jazz Pharmaceuticals, Inc. (“JPI”), Jazz Financing I Limited and Jazz Pharmaceuticals Ireland Limited, as borrowers (collectively with JPI, the “Borrowers”), the lender parties from time to time thereto and Barclays, as administrative agent, collateral agent, letter of credit issuer and swing line lender.

The senior secured credit facility provides for (i) a tranche of term loans made to JPI in the aggregate principal amount of \$557.2 million (“Tranche 1 Term Loans”) and (ii) a revolving credit facility of \$200.0 million. The Tranche 1 Term Loans mature on June 12, 2018 and loans under the revolving credit facility, if any, will mature on June 12, 2017. The senior secured credit facility also provides for one or more incremental term loan facilities and/or increases in commitments under the revolving credit provided that the total aggregate amount of such incremental term loans and increases in the revolving credit facility commitments does not exceed the sum of (A) \$200.0 million and (B) an amount such that at the time of such incurrence and after giving effect thereto on a pro forma basis Parent’s senior secured leverage ratio is less than or equal to 2.75 to 1.

The Tranche 1 Term Loans bear interest, at JPI’s option, at a rate equal to either the LIBOR rate, plus an applicable margin of 2.75% per annum (subject to a 0.75% LIBOR floor), or the prime lending rate, plus an applicable margin equal to 1.75% per annum (subject to a 1.75% prime rate floor). Loans under the revolving credit facility will bear interest, at the applicable Borrower’s option, at a rate equal to either the LIBOR rate, plus an applicable margin of 2.50% per annum, or the prime lending rate, plus an applicable margin equal to 1.50% per annum, subject to reduction by 0.25% or 0.50% based upon Parent’s secured leverage ratio. All overdue amounts will be payable at a rate of 2.00% per annum above the otherwise applicable rate.

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The Borrowers' obligations under the senior secured credit facility and any hedging or cash management obligations entered into with a lender are guaranteed by Parent and each of Parent's existing and subsequently acquired or organized direct and indirect subsidiaries (other than certain immaterial subsidiaries, subsidiaries whose guarantee would result in material adverse tax consequences and subsidiaries whose guarantee is prohibited by applicable law or would result in a violation of violation or breach of, or conflict with, fiduciary duties of such subsidiary's officers, directors or managers). Parent, the Borrowers and such guarantors are collectively as the "Loan Parties." Purchaser is not, and is not currently expected to become, a Loan Party.

The Loan Parties' obligations under the senior secured credit facility are secured, subject to customary permitted liens and other agreed upon exceptions, by a perfected security interest in (a) all tangible and intangible assets of the Loan Parties, except for certain customary excluded assets, and (b) all of the equity interests of the subsidiaries of the Loan Parties held by the Loan Parties (limited, in the case of the equity interests of certain foreign subsidiaries and certain domestic subsidiaries that hold no assets other than equity interests of foreign subsidiaries, to 65% of the voting equity interests of such subsidiaries).

The Borrowers are permitted to make voluntary prepayments of the Tranche 1 Term Loans and any revolving loans at any time without payment of a premium. JPI is required to make mandatory prepayments of Tranche 1 Term Loans (without payment of a premium) with (1) net cash proceeds from certain non-ordinary course asset sales (subject to reinvestment rights and other exceptions), (2) net cash proceeds from issuances of debt (other than certain permitted debt), (3) beginning with the fiscal year ending December 31, 2014, 50% of Parent's excess cash flow (subject to decrease to 25% or 0% if Parent's secured leverage ratio is equal to or less than 2.25 to 1.00 (and greater than 1.25 to 1.00) or 1.25 to 1.00, respectively), and (4) casualty proceeds and condemnation awards (subject to reinvestment rights and other exceptions). The Tranche 1 Term Loans will amortize in equal quarterly installments in an aggregate annual amount equal to 1% of the original principal amount thereof, with any remaining balance payable on the final maturity date of the Tranche 1 Term Loans.

The Credit Agreement contains customary representations and warranties and customary affirmative and negative covenants applicable to Parent and its restricted subsidiaries, including, among other things, restrictions on indebtedness, liens, investments, mergers, dispositions, prepayment of other indebtedness and dividends and other distributions. The Credit Agreement contains a financial covenant that requires Parent to maintain a maximum secured leverage ratio.

### ***Description of the Incremental Term Loans***

Under the commitment letter, JPI would borrow \$500.0 million of Incremental Term Loans under the Credit Agreement. JPI's obligations under the Incremental Term Loans would be guaranteed by Parent and each of the other guarantors under the senior secured credit facility and secured by same assets that secure the obligations under the senior secured credit facility.

It is anticipated that the Incremental Term Loans will mature on June 12, 2018 and will amortize in equal quarterly installments in annual amounts equal to 1.0% of its original principal amount. The applicable interest rate on the Incremental Term Loans is anticipated to be adjusted LIBOR (with a floor of 0.75%) plus 2.75% per annum. Interest based on LIBOR will be payable quarterly in arrears. The definitive financing documentation is also expected to include a base rate option. All overdue amounts will be payable at a rate of 2.00% per annum above the otherwise applicable rate.

It is anticipated that voluntary prepayments of the Incremental Term Loans will be permitted at any time without premium or penalty, except that a 1% premium would apply to a repayment via a repricing of the Incremental Term Loans effected on or prior to the six-month anniversary of the closing date for the Incremental Term Loans. It is further anticipated that the Incremental Term Loans will be subject to mandatory prepayment (without payment of a premium) with (1) net cash proceeds from certain non-ordinary course asset sales (subject to reinvestment rights and other exceptions); (2) net cash proceeds from issuances of debt (other than certain permitted debt); (3) 50% of Parent's consolidated excess cash flow (subject to decrease to 25% or 0% if Parent's

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secured leverage ratio is equal to or less than 2.25 to 1.0 or 1.25 to 1.0, respectively); and (4) casualty proceeds and condemnation awards (subject to reinvestment rights and other exceptions).

The documentation governing the Incremental Term Loans has not been finalized. Accordingly, the terms thereof are subject to change.

### ***Conditions Precedent to the Debt Financing***

The funding of all extensions of credit under Parent's senior secured credit facility, including the Incremental Term Loans and any revolving loans, is subject to the following conditions:

- the accuracy in all material respects of the representations and warranties contained in the loan documents relating to the existing credit facility and absence of any default or event of default thereunder;
- receipt of a notice of borrowing by Barclays for such loans; and
- the absence of any default or event of default that exists or would result from such proposed loans or from the application of the proceeds thereof.

In addition, under the commitment letter, the availability of the Incremental Term Loans is subject to the satisfaction of the following conditions:

- the acquisition, pursuant to the Offer, of 50% of the outstanding Ordinary Shares and ADSs shall have been consummated or will be consummated substantially concurrently with the initial funding of the Incremental Term Loans in accordance with the terms of the Tender Offer Agreement, but without giving effect to any amendments, modifications or waivers by us (other than a "Permitted Minimum Condition Modification" or any such amendment, modification or waiver that is not materially adverse to any interest of the lenders) without the prior written consent of Barclays (it being understood that any reduction in the price that is less than or equal to 10% of the total consideration set forth in the Tender Offer Agreement as of the date of the commitment letter will not be deemed to be materially adverse to the interests of the lenders and will not require the prior written consent of Barclays);
- Barclays having received the financial statements of the Company detailed in Exhibit B to the commitment letter;
- Barclays having been afforded a period of not less than 15 consecutive calendar days following receipt of a customary information memorandum and other customary marketing materials to syndicate the Incremental Term Loans prior to the closing date for the Incremental Term Loans, provided that such period shall not commence earlier than January 6, 2014.
- there not having occurred a company material adverse effect (which is defined in a manner consistent with the Tender Offer Agreement) since June 30, 2013;
- the payment of all fees and expenses required to be paid to the lenders and Barclays;
- execution and delivery of definitive financing documentation for the Incremental Term Loans by each of the loan parties;
- the lenders having received customary opinions of counsel, corporate resolutions, and customary closing documentation and certificates (including a solvency certificate);
- the provision of applicable documentation under applicable "know your customer" and anti-money laundering rules and regulations, including without limitation the Patriot Act; and
- Compliance with the conditions specified under the senior secured credit facility for incremental term loans (including pro forma compliance with the maximum senior secured leverage ratio financial covenant and the limitation on the size of aggregate incremental term loans described above).

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The availability of the debt financing described in the commitment letter may not be considered assured. As of the date hereof, no alternative financing arrangements or alternative financing plans have been made in the event the Incremental Term Loans or revolving loans under the senior secured credit facility are not available at the expiration of the Offer. No plans have been made to finance or repay the Incremental Term Loans or any revolving loans under the senior secured credit facility after the consummation of the transactions contemplated by the Tender Offer Agreement.

### **11. Background of the Offer; Past Contacts or Negotiations with the Company.**

Parent's strategy includes the continued creation of shareholder value by acquiring additional marketed specialty products or products close to regulatory approval to leverage its existing expertise and infrastructure. Thus, Parent regularly evaluates various strategies to improve its competitive position and enhance value for its shareholders, including opportunities for acquisitions of other companies or their assets.

In 2012, Parent initially identified the Company as a potential acquisition candidate due, in part, to the strategic fit with Parent's business and the Company's rights to commercialize Defitelio in Europe. Parent began to actively discuss the Company as a potential acquisition candidate following the Company's announcement, on July 26, 2013, that the European Medicines Agency's Committee for Medicinal Products for Human Use had recommended the granting of a Marketing Authorization Application ("MAA") under exceptional circumstances, for the treatment of severe hepatic veno-occlusive disease ("VOD") in adults and children undergoing hematopoietic stem cell transplantation therapy.

On August 26, 2013, a representative of Parent initiated direct contact by email with Adrian Haigh, Senior Vice President, Commercial Operations of the Company, to learn about the Company's business. During a subsequent call that same day with the same representative of Parent, Mr. Haigh mentioned the Company's potential interest in a strategic transaction with Parent. Mr. Haigh suggested that further information could be shared under a confidentiality agreement. Mr. Haigh sent the representative of Parent non-confidential information concerning the Company and VOD and a draft confidentiality agreement shortly after the call.

On September 4, 2013, Mr. Haigh and the representative of Parent held a call to discuss the Company's strategic goals and the possibility of holding a meeting between the companies' chief executive officers.

On September 5, 2013, a representative of Parent emailed Khalid Islam, Chief Executive Officer of the Company, to make a more formal introduction to Parent and to request a meeting with Dr. Islam.

On September 12, 2013, Dr. Islam and Mr. Haigh met with representatives of Parent in Milan, Italy, including Mr. Iain McGill, the Head of Parent's EUSA International division and Senior Vice President of Parent. At this meeting, the parties discussed various non-confidential aspects of the Company's and Parent's businesses and agreed to explore potential strategic opportunities between the Company and Parent. Dr. Islam indicated that any a decision regarding strategic options for the Company would likely be made in the fourth quarter.

On September 30, 2013, Parent executed and delivered a Confidentiality and Nondisclosure Agreement to the Company, which was countersigned by the Company on October 1, 2013.

On October 3, 2013, Bruce Cozadd, Parent's Chief Executive Officer, and Dr. Islam met in San Francisco, California. During the meeting, the parties discussed potential strategic opportunities between the Company and Parent. Following the meeting, Mr. Cozadd emailed Dr. Islam on October 4, 2013 to communicate Parent's interest in evaluating a potential strategic transaction with the Company.

On October 4, 2013, Parent delivered an initial due diligence request list to the Company.

On October 16, 2013, a representative of Parent spoke with Dr. Islam by phone. During this call, Dr. Islam advised Parent that the Company had engaged Jefferies LLC, ("Jefferies") as its financial advisor and requested a written non-binding proposal from Parent regarding a proposed transaction between Parent and the Company.

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On October 17, 2013, the Company launched an electronic data room and provided representatives of Parent with access to due diligence documents in the data room.

On October 22, 2013, representatives of Parent, including senior managers of its international division, met with the management team of the Company in Como, Italy. Certain other representatives of Parent attended the meeting by phone. A consultant to Parent and representatives of Jefferies also attended the meeting. During the meeting, the Company presented information related to the Company and its business. That same day, representatives of Parent visited the Company's manufacturing plant in Villa Guardia, Italy.

On October 30, 2013, Parent sent a second due diligence request list to the Company.

During the week of November 4, 2013, representatives of Parent and the Company held multiple due diligence calls to cover a wide range of topics, including commercial, technical operations, clinical development, regulatory and intellectual property matters.

On November 14, 2013, Mr. Cozadd called Dr. Islam to advise him that Parent would submit a non-binding proposal to the Company the following week. That same day, the Company launched a new electronic data room and provided access to Parent's representatives.

On November 19, 2013, Mr. Cozadd delivered to Dr. Islam by email a non-binding proposal confirming Parent's interest in acquiring all of the outstanding securities of the Company for a purchase price of U.S. \$53.00 per share in cash and requesting that the Company agree to a 30-day exclusive negotiation period with Parent.

On November 20, 2013, representatives of Barclays, financial advisor to Parent, spoke with Jefferies to discuss Parent's proposal. On behalf of the Company, Jefferies informed Barclays that the Company had commenced a third-party solicitation process for the Company, in which Parent was invited to participate.

On November 21, 2013, Barclays sent Jefferies a high level summary of Parent's confirmatory due diligence requests. From November 21, 2013 through the signing of the definitive agreements on December 19, 2013, Parent and its legal and financial advisors conducted a due diligence investigation of the Company in person, by phone and through document review.

On November 25, 2013, on behalf of the Company, representatives of Jefferies contacted representatives of Barclays and Parent by telephone and confirmed that the Company had commenced a third-party solicitation process and had requested that interested parties submit non-binding proposals by December 20, 2013. Jefferies also indicated that the board of directors of the Company would not be interested in pursuing a transaction at \$53.00 per share, but would likely consider a transaction at a price of \$57.00 per share.

Late in the day on November 26, 2013, Mr. Cozadd and Dr. Islam spoke by phone. Mr. Cozadd indicated that Parent would consider increasing its offer price to \$57.00 per share, subject to confirmatory diligence, in exchange for the Company agreeing to an exclusive negotiating period with Parent and to the inclusion of certain deal protections, including support agreements from certain key shareholders containing appropriate protections for Parent, in connection with the transaction.

On November 27, 2013, Dr. Islam advised Mr. Cozadd by phone that the Company could not agree to an exclusive negotiating period with Parent at that time. Mr. Cozadd and Dr. Islam agreed that the Company would work with Parent to complete Parent's due diligence investigation of the Company and negotiate the terms of the definitive transaction documents.

On November 29, 2013, representatives of the Company, Skadden, Arps, Slate, Meagher & Flom ("Skadden"), legal counsel to the Company and Jefferies held a telephonic meeting with representatives of Parent, Weil, Gotshal & Manges LLP ("Weil"), legal counsel to Parent and Barclays to address a number of



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preliminary considerations relating to a potential transaction between the Company and Parent, including the due diligence process, timing of definitive agreements, and potential antitrust filings.

From November 29, 2013 through the signing of the definitive agreements on December 19, 2013, the parties and their respective legal and financial advisors negotiated the definitive tender offer agreement and support agreements, and Parent negotiated various retention and related arrangements with certain key employees of the Company. During this period, the parties and their respective legal and financial advisors had numerous telephone conferences to discuss the definitive agreements.

On December 4, 2013, Mr. Cozadd had a meeting in New York with representatives of Jefferies that included a discussion of matters related to the potential transaction with the Company.

On December 5, 2013, Mr. Cozadd and Dr. Islam discussed by phone the status of due diligence and agreement negotiations. During the call, Mr. Cozadd requested that the Company accelerate the process that the Company had commenced to seek non-binding proposals from other potential acquirers by December 20, 2013, so that the parties could execute the definitive agreements during the week of December 16, 2013. Later that day, on behalf of the Company, representatives of Jefferies informed representatives of Parent that the Company had determined that if Parent increased its offer price, the Company would consider moving the deadline for submission of non-binding proposals to December 13, 2013.

On December 6, 2013, Mr. Cozadd spoke with Dr. Islam by phone and proposed to increase Parent's offer price to \$57.00 per share in exchange for a commitment from the Company to work towards signing the definitive agreement on or about December 18, 2013 and commencing the tender offer soon thereafter, and the Company's agreement to provide certain deal protections, including support agreements from certain key shareholders containing appropriate protections for Parent, in connection with the transaction.

On each of December 11, December 14, December 16 and December 17, 2013, Mr. Cozadd spoke with Dr. Islam by phone to discuss the status of the agreement negotiation and the due diligence process. On each call, Mr. Cozadd underscored the importance to Parent of receiving support agreements from key shareholders in connection with the transaction.

On December 13, 2013, Weil sent Skadden a form of support agreement for certain shareholders and optionholders of the Company, including certain directors, officers and employees of the Company, expected to be entered into concurrently with the execution of the tender offer agreement. The draft support agreement included a six-month "tail" provision, among other terms.

On December 17, 2013, the Transaction Committee of Parent's board of directors approved the acquisition of the Company by a subsidiary of Parent and related matters.

On December 17, 2013, Parent's legal advisors received written comments from the legal advisors to Sigma-Tau Finanziaria S.p.A. ("Sigma-Tau"), a beneficial owner (together with its affiliated entities) of approximately 17% of the outstanding Ordinary Shares and ADSs that is also represented on the Company Board, to Parent's requested form of support agreement. Later that same day, Parent's and Sigma-Tau's legal advisors negotiated certain terms of the potential support agreement by phone. Sigma-Tau's legal advisors indicated that a business discussion between Parent and Sigma-Tau would be necessary to resolve open business issues related to the support agreement. Later that night, a representative of Parent spoke to an advisor to Sigma-Tau by phone and reiterated Parent's request that Sigma-Tau execute a support agreement. Parent's legal advisors subsequently sent a revised draft of the support agreement by email to Sigma-Tau's legal advisors.

On December 18, 2013, an advisor to Sigma-Tau contacted a representative of Parent by email to indicate that Sigma-Tau had determined not to sign the support agreement requested by Parent.

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On December 18, 2013, Parent was notified that the transaction contemplated by the proposed tender offer agreement had been approved by the board of directors of the Company and that the Company was prepared to enter into the Tender Offer Agreement on the basis that certain shareholders, including certain directors, officers and employees, together representing approximately 23% of the Company's fully diluted share capital, would enter into support agreements with Parent in connection with the transaction.

On December 19, 2013, Parent, Purchaser and the Company executed the Tender Offer Agreement, Parent entered into support agreements with certain of the parties thereto and the Company entered into retention and related arrangements with certain key employees, as requested by Parent. On that same day, Parent and the Company issued a joint press release announcing the execution of the Tender Offer Agreement and the support agreements.

## **12. The Tender Offer Agreement; Other Agreements.**

### **Tender Offer Agreement**

The following is a summary of the material provisions of the Tender Offer Agreement. The following description of the Tender Offer Agreement does not purport to be complete and is qualified in its entirety by reference to the Tender Offer Agreement, a copy of which is filed as Exhibit (d)(1) to the Schedule TO and is incorporated herein by reference. For a complete understanding of the Tender Offer Agreement, you are encouraged to read the full text of the Tender Offer Agreement. The Tender Offer Agreement is not intended to provide you with any other factual information about Parent, Purchaser or the Company. Such information can be found elsewhere in this Offer to Purchase.

The Tender Offer Agreement has been filed herewith as required by applicable SEC regulations and solely to inform investors of its terms. The Tender Offer Agreement contains representations, warranties and covenants, which were made only for the purposes of such agreement and as of specific dates, were made solely for the benefit of the parties to the Tender Offer Agreement, and are intended not as statements of fact, but rather as a way of allocating risk to one of the parties if those statements prove to be inaccurate. In addition, such representations, warranties and covenants may have been qualified by certain disclosures not reflected in the text of the Tender Offer Agreement and may apply standards of materiality in a way that is different from what may be viewed as material by holders of Ordinary Shares and ADSs or other investors in the Company. The holders of Ordinary Shares and ADSs and other investors are not third-party beneficiaries under the Tender Offer Agreement, except to the extent set forth in the Tender Offer Agreement, and should not rely on the representations, warranties and covenants or any descriptions thereof as characterizations of the actual state of facts or conditions of the Company, Parent, Purchaser or any of their respective subsidiaries or affiliates.

*The Offer.* The obligations of Purchaser to, and of Parent to cause Purchaser to, accept for payment and pay for any Ordinary Shares and ADSs validly tendered and not validly withdrawn pursuant to the Offer are subject only to the satisfaction, or waiver by Purchaser or Parent, of the conditions described in Section 14 – “Certain Conditions of the Offer.” Subject to the satisfaction of the Minimum Condition and the other conditions to the Offer, Purchaser will, and Parent will cause Purchaser to, accept for payment and pay for all Ordinary Shares and ADSs validly tendered and not validly withdrawn pursuant to the Offer as promptly as practicable after the expiration of the Offer (as the Expiration Date of the Offer may be extended and re-extended as described below and in compliance with applicable laws) and in any event in compliance with Rule 14e-1(c) under the Exchange Act. The time of such acceptance for payment of Ordinary Shares and ADSs is referred to herein as the “Acceptance Time;” provided that any rights of Purchaser or Parent that arise under the Tender Offer Agreement as a result of the Acceptance Time having occurred will be subject to payment by Purchaser for such Ordinary Shares and ADSs.

The Tender Offer Agreement provides that the Offer Price will be payable in respect of each Ordinary Share and ADS (without duplication for Ordinary Shares underlying the ADSs) validly tendered and not validly withdrawn pursuant to the Offer and will be paid net to the holders of such Ordinary Shares and ADSs in cash,

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without interest, subject to the withholding of any taxes required by applicable law, on the terms and subject to the conditions set forth in the Tender Offer Agreement.

Pursuant to the Tender Offer Agreement, Parent and Purchaser expressly reserve the right to increase the Offer Price, or to make any other changes in the terms and conditions of the Offer, except that the Company's prior written approval is required for Purchaser to (a) decrease the Offer Price, (b) change the form of consideration payable in the Offer, (c) reduce the number of Ordinary Shares and ADSs to be purchased in the Offer, (d) impose conditions to the Offer in addition to the conditions to the offer as set forth in Section 14-"Certain Conditions of the offer", (e) amend, modify or supplement any of the conditions to the Offer in a manner that is adverse to the holders of Ordinary Shares or ADSs, (f) extend the Offer in any manner other than pursuant to certain terms in the Tender Offer Agreement, (g) amend or waive the Minimum Condition (other than a modification to the definition of Minimum Condition, which may be effected at Parent's election and in Parent's sole discretion by delivering written notice of such modification to the Company prior to the Acceptance Time, to replace the reference to "sixty-six and two thirds percent (66  $\frac{2}{3}$ %) of the Adjusted Outstanding Share Number" in such definition with "a majority of the aggregate number of outstanding Company Shares immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying the ADSs)" or (h) amend any terms of the Offer in any manner adverse to the holders of Ordinary Shares and ADSs.

The Offer will initially be scheduled to expire at 12:00 midnight, New York time, on the evening of January 22, 2014.

The Tender Offer Agreement also provides that if, at the initial or at any subsequent Expiration Date of the Offer, any Offer condition is not satisfied or waived by Purchaser or Parent, Purchaser will extend (and re-extend) the Offer and the Expiration Date, to permit such Offer condition to be satisfied, for one or more periods in consecutive increments of up to 20 business days each (or such longer period as Parent and the Company may mutually agree). The length of each such extension may be determined by Parent in its sole discretion. In no event, however, is Purchaser required to extend the Offer beyond April 19, 2014, which is the "Termination Date;" provided, however, that in the event that, on the Termination Date, the only conditions to the Offer that have not been satisfied or waived on or prior to the Termination Date are: (a) the pendency or occurrence of certain governmental actions pending or overtly threatened against Parent, Purchaser or the Company and (b) either or both of (i) the Minimum Condition and (ii) the Company's certification of its compliance with the representations, warranties and covenants made pursuant to the Tender Offer Agreement, then either Parent or the Company may elect to extend the Termination Date, by written notice to the other prior to or on the Termination Date, until September 19, 2014 (the "Extended Termination Date") so long as the conditions set forth in (a) above would be reasonably likely to be satisfied by such Extended Termination Date if the parties use their reasonable best efforts to do so. Pursuant to the Tender Offer Agreement, Purchaser is also required to extend the Offer for any period required by any rule, regulation, interpretation or position of the SEC or its staff or Nasdaq that is applicable to the Offer.

*Subsequent Offering Period.* In addition, the Tender Offer Agreement provides Purchaser will commence a subsequent offering period in accordance with Rule 14d-11 under the Exchange Act. Subject to the terms and conditions of the Tender Offer Agreement and the Offer, Purchaser is required to (and Parent is required to cause Purchaser to) immediately accept for payment, and pay for, all Ordinary Shares and ADSs that are validly tendered pursuant to the Offer during such subsequent offering period.

*Representations and Warranties.* In the Tender Offer Agreement, the Company has made customary representations and warranties to Parent and Purchaser, including representations relating to:

- organization;
- capitalization and indebtedness of the Company;
- subsidiaries;
- authority related to the Tender Offer Agreement and the transactions;

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- authorization;
- non-contravention;
- matters related to certain governmental filings;
- SEC reports and financial statements;
- the internal controls of the Company;
- absence of undisclosed liabilities;
- absence of certain changes;
- compliance with laws;
- litigation;
- employment matters;
- employee plans;
- environmental matters;
- taxes;
- material contracts;
- insurance;
- properties;
- intellectual property;
- the inapplicability of anti-takeover statutes;
- the absence of related party transactions;
- information supplied by the Company;
- broker's or finder's fees payable;
- votes required to be taken prior to consummation of the Offer;
- the opinion of the Company's financial advisor; and
- the Company's compliance with Rule 14d-10 promulgated under the Exchange Act.

In the Tender Offer Agreement, Parent and Purchaser have made customary representations and warranties to the Company, including representations relating to:

- organization;
- authorization;
- non-contravention;
- broker's or finder's fees payable;
- information supplied by Parent or Purchaser;
- litigation applicable to the Offer;
- ownership and prior activities of Purchaser;
- the lack of certain agreements between parent and Purchaser or any officer or director of the Company or any of its Affiliates;
- acknowledgement, taking into account certain assumptions regarding the information publicly supplied by the Company (including accuracy thereof), of the Company's solvency;

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- acknowledgement by Parent and Purchaser that the Company has only made representations and warranties in Article III of the Tender Offer Agreement;
- no interest in Ordinary Shares and ADSs;
- Purchaser and Parent's reliance on only information set forth in the Tender Offer Agreement;
- availability of debt financing (the "debt financing"); and
- intention related to the operations of the Company following the Acceptance Time.

*Operation of the Company's Business.* The Tender Offer Agreement provides that, except as expressly contemplated by the Tender Offer Agreement or as set forth in the confidential disclosure letter, or as consented to in writing by Parent (such consent not to be unreasonably withheld, conditioned or delayed), during the Pre-Closing Period (as defined below), the Company will and will cause its subsidiaries to:

- carry on its business in all material respects in the ordinary course in substantially the same manner as previously conducted and in material compliance with all applicable laws;
- pay its debts and taxes when due, in each case subject to good faith disputes over such debts or taxes contested in good faith by any appropriate action and for which adequate reserves are established to the extent required by GAAP (as defined in the Tender Offer Agreement);
- use commercially reasonable efforts to:
  - preserve intact its present business organization;
  - pay or perform all material obligations when due, subject to good faith disputes over such obligations;
  - keep available the services of its present officers and employees; and
  - preserve its relationships with material customers, suppliers, investigators, distributors, licensors, licensees and others with which it has material business dealings.
- use its reasonable best efforts to prepare and prosecute the approval by the United States Food and Drug Administration of the Defibrotide NDA consistent with past practice and to develop Defibrotide;
- use its reasonable best efforts to obtain and maintain on a commercially reasonable basis quantities of finished Defibrotide (as defined in the Tender Offer Agreement) drug product and related raw materials and components that the Company reasonably expects to be required for the anticipated commercial launch of Defibrotide; and
- use commercially reasonable efforts to (i) preserve and maintain the validity of all authorizations for the manufacture of and marketing of medicinal products and orphan designations for medicinal products granted by any Regulatory Authority to the Company or any of its subsidiaries and (ii) ensure that such authorizations or orphan designations are not varied, suspended or withdrawn.

As defined in the Tender Offer Agreement, "Pre-Closing Period" means the date on which the parties entered into the Tender Offer Agreement through the earlier to occur of the termination of the Tender Offer Agreement and the Acceptance Time.

In addition, the Tender Offer Agreement provides that, except as expressly contemplated or permitted by the Tender Offer Agreement, as set forth in the confidential disclosure letter, or as approved in advance by Parent in writing (such approval not to be unreasonably withheld, conditioned or delayed with respect to certain matters), during the Pre-Closing Period, the Company will not, and will not permit its subsidiaries to, do any of the following:

- declare, set aside or pay any dividends on, or make any other distributions (whether in cash, stock or property) in respect of, any of its capital stock or other equity interests, except for dividends by a wholly owned subsidiary of the Company to the Company;

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- except in connection with outstanding options to purchase Ordinary Shares or ADSs pursuant to the Company's stock option plans ("Company Options"), purchase, redeem or otherwise acquire shares of capital stock or other equity interests of the Company or its subsidiaries or any Company Options, warrants, or rights to acquire any such shares or other equity interests;
- split, combine, reclassify or otherwise amend the terms of any of its capital stock or other equity interests;
- issue, sell, deliver or agree or commit to issue, sell or deliver any shares of capital stock or any securities of the Company or any of its subsidiaries, except with respect to the issuance of Ordinary Shares or ADSs of the Company related to outstanding Company Options;
- adopt, amend, authorize or propose to amend its articles of incorporation or bylaws (or similar organizational documents) except for amendments as required by law in connection with the exercise of any outstanding Company Options;
- create or form any subsidiary;
- acquire or agree to acquire any entity other than through a cash purchase not to exceed \$1,000,000, and such acquisitions may not, in the aggregate, exceed \$2,500,000;
- acquire or agree to acquire any assets that are otherwise material to the Company and its subsidiaries, other than in the ordinary course of business consistent with past practice;
- directly or indirectly sell, lease, license, sell and leaseback, abandon, mortgage or otherwise encumber or subject to any lien or otherwise dispose of any of its material properties, assets or rights or any interest therein, except certain non-exclusive licenses in the ordinary course of business, sales of inventory of API and company products, and equipment and property no longer used in the operation of business;
- propose or adopt a plan of complete or partial liquidation, dissolution, merger, consolidation, restructuring, recapitalization or other reorganization of the Company or any of its subsidiaries, except for the transactions contemplated by the Tender Offer Agreement;
- incur, create, assume or otherwise become liable for any specified forms of indebtedness;
- amend, modify or refinance any indebtedness;
- make any loans, advances or capital contributions to, or investments in any person or entity other than the Company or any of its subsidiaries, with certain exceptions;
- syndicate or issue, or attempt to syndicate or issue or announce the syndication or issuance of any debt facility or debt security that competes with Parent's debt financing activities in connection with the Offer (including any renewal or re-debt financing of any of the Company's or its subsidiaries' existing debt facilities or securities);
- incur or commit to incur any capital expenditure or authorization or commitment with respect thereto in excess of \$500,000 individually or \$1,000,000 in the aggregate, with certain exceptions;
- pay, discharge, settle or satisfy any claims, liabilities or obligations (whether absolute, accrued, asserted or unasserted, contingent or otherwise), in an excess \$500,000 individually or \$1,000,000 in the aggregate (with certain exceptions);
- cancel any material indebtedness;
- waive, release, grant or transfer any right of material value;
- compromise, settle or agree to settle any claim or action in excess of \$500,000 individually or \$1,000,000 in the aggregate, and in no case will such compromise, settlement or agreement impose any equitable relief, or the admission of wrongdoing, on the Company;
- change its financial or tax accounting methods, principles or practices, except as required by a change in the GAAP or applicable law;

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- take certain actions related to Company's taxes;
- change its fiscal year;
- take certain actions related to current or former directors, officers, employees or independent contractors of the Company or any of its subsidiaries or affiliates, with certain exceptions;
- fail to use commercially reasonable efforts to keep in force insurance policies or replacement or revised provisions regarding insurance coverage with respect to material assets, operations and activities of the Company and its subsidiaries as currently in effect;
- renew or enter into certain non-compete, exclusivity or similar agreements;
- enter into any new material lease of real property or materially amend the terms of any existing material lease of real property, other than in the ordinary course of business;
- forgive any loans to any employees, officers or directors of the Company and its subsidiaries, or any of their respective affiliates or associates;
- take certain actions related to the Company's employee benefits plans;
- enter into or become bound by, or permit any of the assets owned or used by the Company to become bound by, any material contract, or seek to amend, terminate or waive, or exercise any material right or remedy under, any material contract or enter into or become bound by, or seek to amend, terminate or waive, or exercise any material right or remedy under, any contract pursuant to which the Company or any of its subsidiaries grants or has granted to any person or entity an exclusive license to practice or exploit the material intellectual property rights of the Company;
- other than in the ordinary course of business consistent with past practices or as required by GAAP, write off as uncollectible, or establish any extraordinary reserve with respect to, any receivable or other indebtedness;
- take any actions that may undermine validity of or the Company's ability to transfer any authorization for the manufacturing or marketing of medicinal products or any orphan designation for a medicinal product to Parent or Purchaser;
- enter into or become bound by any distribution agreement or commercial tendering contract;
- grant any exclusive rights with respect to, or divest any of, the Company's intellectual property, other than in the ordinary course of business consistent with past practices; or
- authorize any of, or commit, resolve or agree to take any of, the foregoing actions unless approved in advance by Parent in writing (such approval not to be unreasonably withheld, conditioned or delayed for certain of the foregoing provisions).

*No Solicitation by the Company; Other Offers.* The Tender Offer Agreement provides that during the Pre-Closing period, the Company will not and will cause its subsidiaries and officers, directors, employees, investment bankers, attorneys, accountants, consultants and other agents and advisors, not to: (a) solicit, initiate or knowingly encourage (including by way of providing non-public information) the submission or announcement of any inquiries, proposals or offers that constitute or would reasonably be expected to result in an Acquisition Proposal (as defined below); (b) provide any non-public information concerning the Company or any of its subsidiaries to any person or entity in connection with or in response to an Acquisition Proposal; (c) engage in any discussions or negotiations with any person or entity with respect to any Acquisition Proposal; (d) approve, adopt, endorse or recommend any Acquisition Proposal; or (e) enter into any agreement, letter of intent or similar document relating to any Acquisition Transaction (as defined below) or requiring the Company to cause restrictions contained in any state takeover or similar laws not to apply to a third party or any Acquisition Proposal.

Notwithstanding the foregoing, if at any time prior to the Acceptance Time, (a) the Company has received an unsolicited, bona fide written Acquisition Proposal (other than an Acquisition Proposal that arises as a result

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of a breach of any of the provisions set forth in the immediately preceding paragraph) that is reasonably likely to result in a Superior Proposal (as defined below), (b) the Company Board determines in good faith, after having consulted with outside legal counsel, that the failure to take such action would be inconsistent with its fiduciary obligations to the Company shareholders and other stakeholders of the Company under applicable laws, (c) prior to furnishing or making available any non-public information to, or entering into discussions or negotiations with, such person or entity, the Company gives Parent written notice of the identity of the person or entity making the Acquisition Proposal and of the Company's intention to furnish or make available non-public information to, or enter into discussions or negotiations with, such person or entity, and the Company receives from such person or entity an executed confidentiality agreement containing provisions (including nondisclosure provisions and use restrictions) at least as favorable to the Company as the provisions of the Confidentiality Agreement (as defined under "Other Agreements – Confidentiality Agreement"), and (d) simultaneously with furnishing or making available any non-public information to such person or entity, the Company makes available such non-public information to Parent (to the extent the Company has not previously made available such non-public information to Parent), then the Company may (i) furnish information with respect to the Company and its subsidiaries to the person or entity making such Acquisition Proposal and (ii) engage in discussions or negotiations with the person or entity making such Acquisition Proposal regarding such Acquisition Proposal.

The Company will promptly (and, in any event, within one business day) notify Parent in the event that the Company, any subsidiary of the Company or any of their respective representatives receives any Acquisition Proposal, including the identity of the person or entity making such Acquisition Proposal, and the material terms and conditions thereof and, if applicable, provide Parent with copies of any written requests, proposals or offers from the person or entity making such Acquisition Proposal promptly after receipt thereof. The Company is required to keep Parent informed with respect to: (a) the status of any such Acquisition Proposal and (b) the status and material terms of any material modification thereto.

The Tender Offer Agreement provides that, as of the date of the Tender Offer Agreement, the Company will, and will cause each of its subsidiaries and its and their representatives to, immediately cease and cause to be terminated any solicitations or discussions with any other person or entity with respect to any actual or potential Acquisition Proposal.

The Tender Offer Agreement provides that the Company agrees not to release or permit the release of any person or entity from, or to waive or permit the waiver of any provision of, any confidentiality, non-solicitation, no hire, "standstill" or similar contract to which the Company is a party or under which the Company has any rights to the extent permitted thereunder, and will use commercially reasonable efforts to cause each such agreement to be enforced (it being understood that any automatic termination of any such provision resulting from the execution and delivery of the Tender Offer Agreement will not be considered a breach of the foregoing). The Company also is to promptly request that each person or entity that has executed a confidentiality or similar agreement with the Company or its subsidiaries within the eighteen months prior to the date of the Tender Offer Agreement in connection with its consideration of a possible Acquisition Transaction or a possible equity investment in the Company return to the Company or destroy all confidential information heretofore furnished to such person or entity by or on behalf of the Company.

As defined in the Tender Offer Agreement, "Acquisition Proposal" means any inquiry of or communication to the Company or any offer or proposal, or indication of interest in making an offer or proposal (other than an offer, proposal or indication of interest in making an offer or proposal by Parent or Purchaser), in each case, relating to any Acquisition Transaction.

As defined in the Tender Offer Agreement, "Acquisition Transaction" means any transaction or series of related transactions (other than the transactions contemplated by the Tender Offer Agreement) involving: (a) any acquisition or purchase from the Company or any of its subsidiaries by any person or entity or "group" (as defined in or under Section 13(d) of the Exchange Act), directly or indirectly, of more than a 15% interest in the



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total outstanding voting securities of the Company or any of its subsidiaries; (b) any tender offer (including self-tender) or exchange offer that if consummated would result in any person or entity or “group” (as defined in or under Section 13(d) of the Exchange Act) beneficially owning 15% or more of the total outstanding voting securities of the Company or any of its subsidiaries; (c) any merger, consolidation, business combination, share exchange, issuance of securities, acquisition of securities, recapitalization or other similar transaction involving the Company or any of its subsidiaries, the business(es) of which, individually or in the aggregate, constitute more than 15% of the assets of the Company and its subsidiaries, taken as a whole, pursuant to which the shareholders of the Company or such subsidiary immediately preceding such transaction hold less than 85% of the voting equity interests in the surviving or resulting entity of such transaction or any parent entity thereof; (d) any sale, lease, exchange, transfer, license or disposition (in each case, other than in the ordinary course of business) of more than 15% of the assets of the Company and its subsidiaries, taken as a whole (measured by the lesser of book or fair market value thereof); or (e) any combination of the foregoing.

As defined in the Tender Offer Agreement, “Superior Proposal” means any bona fide written Acquisition Proposal involving an Acquisition Transaction that, if consummated, would result in a person, entity or “group” (as defined in or under Section 13(d) of the Exchange Act) owning more than fifty percent (50%) of the outstanding voting securities of the Company or more than fifty percent (50%) of the assets of the Company and its subsidiaries, taken as a whole, which the Company Board will have determined in good faith (after consultation with its financial advisor and its outside legal counsel) is more favorable to the holders of Ordinary Shares and ADSs (in their capacity as such) and other stakeholders of the Company than the transactions contemplated by the Tender Offer Agreement.

*Change in Company Board Recommendation.* The Tender Offer Agreement provides that neither the Company Board nor any committee thereof will (a) withdraw (or modify in a manner adverse to Parent or Purchaser), or propose (other than proposals made at any meeting of the Company Board solely among the participants at such meeting) or adopt any resolution to withdraw (or modify in a manner adverse to Parent or Purchaser), the Company Board Recommendation, (as defined in the Tender Offer Agreement) (it being understood that the Company Board Recommendation will be deemed to have been modified in a manner adverse to Parent or Purchaser if it will no longer be by the unanimous vote of all directors of the Company, other than those directors that have deemed themselves conflicted (any action described in clause (a) being referred to herein as a “Company Board Recommendation Change”), (b) resolve in favor of or recommend publicly or to any third party the adoption of any Acquisition Proposal, or (iii) cause or permit the Company or any of its subsidiaries to execute or enter into any letter of intent, memorandum of understanding, agreement in principle, merger agreement, acquisition agreement or other similar agreement providing for any Acquisition Proposal, other than any confidentiality agreement referred to in Section 6.1(b) of the Tender Offer Agreement (an “Acquisition Agreement”).

Notwithstanding anything to the contrary contained in the Tender Offer Agreement, at any time prior to the Acceptance Time, the Company Board may effect a Company Board Recommendation Change if: (a) an unsolicited, bona fide, written Acquisition Proposal that did not otherwise result from a breach of any provision of the Tender Offer Agreement or any “standstill” or similar agreement or provision under which the Company or any of its subsidiaries has any rights is made to the Company and is not withdrawn, (b) the Company provides to Parent written notice at least 24 hours (or simultaneous with the Company Board if lesser notice is given to the Company Board) before any meeting of the Company Board at which the Company Board will consider the possibility of withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change in connection with such Acquisition Proposal, which notice specifies the reasons for holding such meeting; (c) at such meeting, the Company Board determines in good faith (after consulting with the Company’s financial advisor and outside legal counsel) that such Acquisition Proposal constitutes a Superior Proposal, (d) at such meeting, the Company Board determines in good faith, after consulting with its outside legal counsel, that, in light of such Superior Proposal, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable law, (e) the Company promptly notifies Parent, in writing, at least four business days before withdrawing the Company Board Recommendation or

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otherwise making any Company Board Recommendation Change: (i) that it has received a Superior Proposal that did not result from a breach of its non-solicitation obligations; (ii) that the Company Board intends to withdraw the Company Board Recommendation or otherwise make a Company Board Recommendation Change; and (iii) specifying the material terms and conditions of such Superior Proposal, including the identity of the person or entity making such Superior Proposal (and attaching the current version of any written agreement relating thereto) (it being understood and agreed that any change to the consideration payable in connection with such Superior Proposal or any other material modification thereto will require a new advance written notice by the Company (except that the four business day period referred to above will be reduced to two business days)), (f) during such notice period, if requested by Parent, the Company engages in good faith negotiations with respect to any proposal made by Parent to amend the Tender Offer Agreement in such a manner that obviates the need for withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change, and (g) at the time of any Company Board Recommendation Change, the Company Board confirms the determinations referred to in clauses (c) and (d) above (taking into account any changes to the terms of the Tender Offer Agreement proposed by Parent as a result of the negotiations required by clause (f) above or otherwise). The Company will notify Parent promptly upon any withdrawal of the Company Board Recommendation or other Company Board Recommendation Change.

Notwithstanding anything to the contrary contained in the Tender Offer Agreement, at any time prior to the Acceptance Time, the Company Board may effect a Company Board Recommendation Change if: (a) there occurs or arises after the date of the Tender Offer Agreement a material event, material development or material change in circumstances that does not relate to any Acquisition Proposal that was not known by or, if known, the consequences of which (i) were not known by the Company or any of its subsidiaries on the Agreement Date, and (ii) would not have been reasonably expected to have occurred, which event, development or change in circumstance, or any material consequences thereof, becomes known to the Company Board prior to the Acceptance Time (any such material event, material development or material change in circumstances unrelated to an Acquisition Proposal being referred to as an "Intervening Event"); (b) the Company provides to Parent written notice at least 24 hours (or simultaneous with the Company Board if lesser notice is given to the Company Board) before any meeting of the Company Board at which the Company Board will consider the possibility of withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change in connection with such Intervening Event, which written notice specifies the reasons for holding such meeting and a reasonably detailed description of such Intervening Event; (c) at such meeting, the Company Board determines in good faith, after consulting with the Company's outside legal counsel, that, in light of such Intervening Event, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable law; (d) the Company promptly notifies Parent, in writing, at least four business days before withdrawing the Company Board Recommendation or otherwise making any Company Board Recommendation Change that the Company Board has determined that, in light of such Intervening Event, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable law and that it intends to withdraw the Company Board Recommendation or otherwise make a Company Board Recommendation Change as a result of such Intervening Event; (e) during such notice period, if requested by Parent, the Company engages in good faith negotiations with respect to any proposal made by Parent to amend the Tender Offer Agreement in such a manner that obviates the need for withdrawing the Company Board Recommendation or otherwise effecting a Company Board Recommendation Change; and (f) at the time of any Company Board Recommendation Change, the Company Board confirms its determination that, in light of such Intervening Event, the failure to withdraw the Company Board Recommendation or otherwise effect a Company Board Recommendation Change would be inconsistent with its fiduciary obligations to the Company Shareholders and other stakeholders of the Company under applicable law (taking into account any changes to the terms of the Tender Offer Agreement proposed by Parent).

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Nothing described above under “No Solicitation by the Company; Other Offers” and “Change in Company Board Recommendation” or elsewhere in the Tender Offer Agreement will prohibit the Company from (a) taking and disclosing to its shareholders a position contemplated by Rule 14d-9 or Rule 14e-2(a) promulgated under the Exchange Act or Item 1012(a) of Regulation M-A under the Exchange Act, or (b) making any “stop-look-and-listen” communication to the Company Shareholders pursuant to Section 14d-9(f) promulgated under the Exchange Act (or any similar communications to the Company Shareholders), in the case of each of clauses (a) and (b), in which the Company indicates that it has not made a Company Board Recommendation Change; provided, however, that these exceptions do not broaden the extent to which a Company Board Recommendation Change will be permitted under the Tender Offer Agreement.

*Reasonable Best Efforts.* The Tender Offer Agreement provides that each party is required to use its reasonable best efforts to take, or cause to be taken, all actions, and to do, or cause to be done, and to assist and cooperate with the other parties in doing, all things reasonably necessary, proper or advisable to consummate and make effective, in the most expeditious manner practicable, the transactions contemplated by the Tender Offer Agreement, including using their reasonable best efforts to: (a) cause the conditions of the Offer to be satisfied or fulfilled; and (b) obtain all necessary actions or material non-actions, waivers, consents, approvals, orders and authorizations from governmental entities, the expiration or termination of any applicable waiting periods, making all necessary registrations, declarations and filings (including registrations, declarations and filings with governmental entities, if any).

*Access.* Throughout the Pre-Closing Period, the Company is to allow Parent and its accountants, legal counsel and other representatives (including Parent’s debt financing sources or prospective debt financing sources and their respective advisors and representatives) reasonable access during normal business hours, upon reasonable notice, to the properties, books and records and personnel of the Company to enable Parent to obtain all information concerning the business, as Parent may reasonably request and only in accordance the terms and conditions of the Confidentiality Agreement (as defined under “Other Agreements – Confidentiality Agreement”). Notwithstanding anything to the contrary in the previous sentence, the Company will not be required to provide access to, or to disclose information, where such access or disclosure would reasonably be expected to jeopardize the attorney-client privilege (or similar protections) of the Company or its subsidiaries or contravene any applicable law or contract, but in such circumstance, the Company will cooperate with Parent to implement a procedure to permit access to or disclosure of such information in a manner that would not jeopardize attorney-client privilege (or similar protections) or contravene such applicable law or contract.

*Debt Financing.* Each of Parent and Purchaser will:

- not permit any amendment or modification to be made to, or any waiver of any provision or remedy pursuant to, Parent’s commitment letter with Barclays if such amendment, modification or waiver would or would reasonably be expected to delay or prevent in any material respect the ability of Purchaser to consummate the Offer; and
- unless financing for the Offer has otherwise been obtained, keep the Company informed on a reasonable basis and in reasonable detail of the status of its efforts to arrange the debt financing and use commercially reasonable efforts to maintain in effect the commitment letter (as described more fully in Section 10 – “Source and Amount of Funds”).

Notwithstanding the foregoing, neither Parent nor Purchaser will be required to commence, participate in, pursue or defend any claim or action against or involving any debt financing sources. In the event that any portion of the debt financing becomes unavailable on the terms and conditions contemplated in the commitment letter or otherwise reasonably acceptable to the Parent, each of Parent and Purchaser will use commercially reasonable efforts to obtain alternative financing promptly following the occurrence of such event. Parent will deliver to the Company a copy of any amendment, modification, waiver or supplement to the commitment letter that would reasonably be expected to materially impact the availability of the debt financing or materially decrease the amount of financing that could be expected to be provided under the commitment letter as promptly

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as practicable after the execution thereof; provided that any such amendments, modification, waiver or supplement may be redacted to the extent requested by the applicable debt financing source. Parent and Purchaser acknowledged and agreed that their obligations under the Tender Offer Agreement, including their obligations to consummate the Offer, are not subject to, or conditioned on, receipt of the debt financing or any other financing.

*Cooperation by the Company.* During the Pre-Closing Period, the Company will use commercially reasonable efforts to provide Parent with all cooperation reasonably requested by Parent in connection with Parent's financing of the Offer, including by (a) providing to Parent information reasonably requested by Parent required for purposes of the financing and participate and make senior management of the Company available in meetings to discuss any such information, (b) preparing and providing to Parent financial statements and projections (including providing audited financial statements for the year ending December 31, 2013 no later than March 31, 2014) reasonably requested by Parent for purposes of the financing, (c) assisting in the preparation of those sections of any customary offering memoranda and other customary presentation materials, private placement memoranda, prospectuses and similar documents (including, if applicable, the delivery of one or more customary representation letters) that relate to the Company and its business, and (d) obtaining the consent of, and customary comfort letters from, Reconta Ernst & Young S.p.A. (including by providing customary management letters and requesting legal letters to obtain such consent) if necessary or desirable for Parent's use of the Company's financial statements. However, the Tender Offer Agreement does not require such cooperation to the extent it would (i) unreasonably disrupt the conduct of the business or operations of the Company or its subsidiaries (in the reasonable judgment of the Company), (ii) require the Company or any of its subsidiaries to agree to pay any fees, reimburse any expenses or otherwise incur any liability or give any indemnities prior to the Acceptance Time for which it is not promptly reimbursed or simultaneously indemnified or (iii) require the Company or any of its subsidiaries to take any action that would reasonably be expected to conflict with, or result in any violation or breach of, or default under, the Company's articles of association, the Company's bylaws (*Statuto*), any applicable laws, or any contract. Parent may reasonably use logos of the Company solely in connection with the debt financing of the Offer; provided that (A) such logos are used solely in a manner that is not intended to nor reasonably likely to harm or disparage any of the Company and its subsidiaries or the reputation or goodwill of any of the Company and its subsidiaries and its or their intellectual property rights, (B) the logos will be used in accordance with any written guidelines provided by the Company to Parent, and (C) Parent must immediately terminate use of such logos upon termination of the Tender Offer Agreement, and provided further that the Company will have an opportunity to review and reasonably object to any use of the Company's and its subsidiaries' logos prior to the dissemination of any materials in which such logos are used. Any goodwill resulting from any such use of the logos will inure to the benefit of the Company or its subsidiaries, as applicable.

*Financing Sources.* Notwithstanding anything to the contrary contained in the Tender Offer Agreement, the Company will not bring or support any legal action, suit or proceeding (whether at law, in equity, in contract, in tort or otherwise) against any financing source in any way relating to the Tender Offer Agreement or any of the transactions contemplated by the Tender Offer Agreement (including the debt financing), including any dispute arising out of or relating in any way to the debt financing or the performance thereof.

*Directors' and Officers' Indemnification and Insurance.* For five years after the Acceptance Time (the "Indemnification Period"), Parent will cause the Company and its subsidiaries and successors to honor and fulfill the obligations of the Company and its subsidiaries under its indemnification agreements in effect as of the date of the Tender Offer Agreement with any of the Company's or its subsidiaries' respective current or former directors and officers and any person who becomes a director or officer of the Company or any of its subsidiaries prior to the Acceptance Time (the "Indemnified Parties") with respect to actions taken by such Indemnified Party in his or her capacity as a director or officer of the Company or its subsidiary. In the event that any claim or claims are asserted or made within the Indemnification Period, all rights to indemnification in respect of any such claim or claims will continue until disposition of any and all such claims.

Further, during the Indemnification Period, Parent and the Company will maintain in effect the Company's current directors' and officers' liability insurance ("D&O Insurance") in respect of acts or omissions occurring at

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or prior to the Acceptance Time, covering each person covered by the D&O Insurance immediately prior to the Acceptance Time, on terms no less favorable, in the aggregate, than those of the D&O Insurance in effect on the date the Tender Offer Agreement is executed and with an insurance company with the same or better credit rating as the insurance company providing the current D&O Insurance.

The Company may, at its option, substitute policies of Parent, Purchaser, the Company or any of their respective subsidiaries containing certain terms with respect to coverage, retention, limitations of liability and amounts no less favorable, in the aggregate, to such persons than the D&O Insurance, or request that the Company obtain such extended reporting period coverage under its existing insurance program (to be effective as of the Acceptance Time). Parent or Purchaser will not be obligated to pay annual premiums in excess of 250% of the annual premium for coverage under the D&O Insurance in effect on the date of the Tender Offer Agreement (such 250% amount, the “Maximum Annual Premium”), and if the annual premiums of such insurance coverage exceed such amount, Parent, Purchaser and the Company will be obligated to obtain a policy with the greatest coverage available for a cost not exceeding the Maximum Annual Premium. Parent, Purchaser or the Company may prior to the Acceptance Time, purchase a five-year “tail” prepaid policy on the D&O Insurance containing certain terms no less favorable, in the aggregate, than the D&O Insurance in effect on the date of the Tender Offer Agreement and with an insurance company with the same or better credit rating as the insurance company providing the current D&O Insurance. In the event that Parent, Purchaser or the Company purchases such a “tail” policy prior to the Acceptance Time, Parent, Purchaser and the Company, as applicable, will maintain such “tail” policy in full force and effect through such five-year period, in lieu of all other obligations of Parent, Purchaser and the Company with respect to the D&O Insurance.

The Tender Offer Agreement provides that if Purchaser or the Company or any of its successors or assigns consolidates with or merges into any other person or entity and will not be the continuing or surviving corporation or entity of such consolidation or merger or transfers all or substantially all of its properties and assets to any person or entity, then, and in each such case, proper provisions will be made so that the successors and assigns of the Company will assume all of the obligations of Parent, Purchaser and the Company related to the rights and obligations described under “Directors’ and Officers’ Indemnification Insurance.”

The obligations described under “Directors’ and Officers’ Indemnification Insurance” may not be terminated, amended or otherwise modified in such a manner as to adversely affect any Indemnified Party (or certain other persons) without the prior written consent of such affected Indemnified Party or other person who is a beneficiary under the D&O Insurance or the “tail” policy. The rights of the Indemnified Parties (and certain other persons) are in addition to, and not in substitution for, any other rights that such persons may have under the certificate or articles of incorporation, bylaws or other equivalent organizational documents, any and all indemnification agreements of or entered into by the Company or any of its subsidiaries, or applicable law (whether at law or in equity).

*Rule 14d-10 Matters; Employment Compensation Approval.* Prior to the expiration of the Offer, the Company will take all such steps as may be required to cause any employment compensation, severance or employee benefit arrangements that have been or will be entered into after the date of execution of the Tender Offer Agreement by the Company or its subsidiaries with current or future directors, officers or employees of the Company or its subsidiaries to be exempt under Rule 14d-10(d) under the Exchange Act and to ensure that any such arrangements fall within the safe harbor provisions of such rule.

*Shareholder Litigation.* The Tender Offer Agreement provides that the Company must promptly advise Parent in writing of any litigation commenced after the date of the Tender Offer Agreement against the Company or any of its directors by any shareholders of the Company (on their own behalf or on behalf of the Company), relating to the Tender Offer Agreement or the transactions contemplated thereby and will keep Parent reasonably informed regarding any such litigation. The Company will give Parent the opportunity to consult with the Company regarding the defense or settlement of any such shareholder litigation and will consider in good faith Parent’s views with respect to such litigation and will not settle any such litigation without the prior written consent of Parent (which consent will not be unreasonably withheld, conditioned or delayed).

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*Public Disclosure.* Parent and the Company will consult with each other before issuing any press releases or otherwise making any public statements with respect to the Tender Offer Agreement and the transactions contemplated thereby, and will not issue any such press release or make any such public statement prior to such consultation, except as may be required by applicable law or any listing agreement with a national securities exchange, in which case commercially reasonable efforts to consult with the other party will be made prior to any such release or public statement. Nonetheless, (a) each party may, without such consultation or consent, make any public statement in response to questions from the press, analysts, investors or those attending industry conferences and make internal announcements to employees, so long as such statements are consistent with previous press releases, public disclosures or public statements made jointly by the parties (or individually, if approved by the other party), and (b) the Company will not be required to consult or agree with Parent in connection with any public disclosure, including issuing any factually accurate public statement by the Company that describes the Company's receipt of an Acquisition Proposal and the operation of the Tender Offer Agreement with respect thereto or "stop-look-and listen communication" pursuant to, or otherwise complying with, the provisions of Rule 14d-9 promulgated under the Exchange Act or any similar communication to the Company's shareholders, in connection with any Acquisition Proposal or Superior Proposal, with certain exceptions. The Company will consult with Parent and consider the views and comments of Parent before the Company, any subsidiary of the Company or any of their respective Representatives sends any emails or other documents to the employees of the Company generally or otherwise communicates with such employees generally, with respect to the Offer or any of the other transactions contemplated by the Tender Offer Agreement.

*Treatment of the Company's Equity Awards.* As soon as practicable following the date of the Tender Offer Agreement (and, in any event, prior to the Acceptance Time), the Company Board will adopt resolutions and take such other actions as may be required to (i) provide that all outstanding and unvested Company Options will become fully vested immediately prior to the Acceptance Time, and (ii) provide notice to holders of Company Options that all Company Options that are outstanding as of the Acceptance Time may be exercised for Ordinary Shares immediately following the Acceptance Time and, prior to the expiration date of the subsequent offering period with respect to the Offer, as it may be extended, such holders may tender such Ordinary Shares in exchange for the Offer Price pursuant to the Offer, as extended by such subsequent offering period, and the terms and conditions set forth in the Tender Offer Agreement.

As soon as practicable following the date of the Tender Offer Agreement, the Company will provide notice to each holder of outstanding Company Options (except directors, consultants, independent contractors, members of senior management or any executive officer of the Company as defined under Section 13(k) of the Exchange Act or Italian law or any officer of the Company or a subsidiary for whom such an arrangement would violate any applicable law) that such holder may elect to pay the aggregate exercise price for the exercise of such holder's outstanding vested Company Options with the funds made available by the Company (on terms and conditions that conform to applicable law) in consideration for such holder's irrevocable instructions to the Company or its broker to (i) exercise such Company Options for Ordinary Shares after the Acceptance Time and, (ii) prior to the expiration date of the subsequent offering period with respect to the Offer, as it may be extended, tender each such Ordinary Share in exchange for the Offer Price pursuant to the Offer, as extended by such subsequent offering period, and the terms and conditions set forth in the Tender Offer Agreement.

The Company and its subsidiaries are required to comply with applicable law requiring withholding, reporting or remittance of any Taxes due with respect to any Company Options that are exercised following the Acceptance Time.

*Employee Matters.* The Company will, within fifteen days following the date of the Tender Offer Agreement, deliver to Parent an accurate and complete list as of the date of the Tender Offer Agreement of all employees and certain independent contractors or consultants currently providing services to the Company or any of its subsidiaries and indicating certain employment information about each such person.

Unless a higher protection is given to the continuing employees (as defined below) of the Company under applicable law or any such continuing employee of the Company agrees otherwise, for a period of at least twelve

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months following the Acceptance Time, Parent will, and will cause its subsidiaries to, provide to all continuing employees of the Company located outside of the United States compensation and benefits (not including Company Options or other equity compensation, retention or bonuses, with certain exceptions) that are, in the aggregate, no less favorable than the compensation and benefits being provided to continuing employees of the Company located outside of the United States immediately prior to the Acceptance Time.

Parent will cause its subsidiaries to honor in accordance with their terms as in effect immediately prior to the Acceptance Time all existing employment, change of control, severance and retention arrangements between the Company or any of its subsidiaries, on the one hand, and any current or former employee, director or consultant of the Company or any of its subsidiaries, on the other hand, and will recognize for such purposes continuous service with the Company or its subsidiaries, including predecessor employers, of participating continuing employees of the Company under such plans.

Nothing described above under “Employee Matters” or elsewhere in the Tender Offer Agreement prohibits Purchaser or its subsidiaries from amending or terminating any employment plans, programs, agreements or arrangements, so long as such amendment or termination complies with the terms of any such plans, programs, agreements or arrangements and applicable law, including specifically obtaining any necessary or required consents.

Furthermore, the terms and conditions of employment after the Acceptance Time for continuing employees of the Company whose employment is subject to a collective bargaining agreement, works council agreement or similar contract with any labor organization, works council or employee association will be governed by such agreement or contract until its expiration, modification or termination in accordance with its terms and applicable law.

Unless otherwise requested by Parent in writing at least five days prior to the Acceptance Time, the Company has agreed to take (or cause to be taken) all actions necessary or appropriate to terminate, effective as of the day immediately prior to, and conditioned upon, the Acceptance Time, any Employee Benefit Plan that is intended to be qualified under Section 401(k) of the Code (the “401(k) Plan”) and, prior to and conditioned upon such termination, fully vest any and all unvested amounts of the accounts of all individuals who are participants under the 401(k) Plan at the time of such termination. The Company is to deliver to Parent an executed copy of such resolutions as soon as practicable following adoption of such resolutions and to fully comply with such resolutions.

As defined in the Tender Offer Agreement “continuing employees” of the Company means all employees of the Company (a) who are offered and timely accept employment by Parent or any subsidiary of Parent, (b) who continue their employment with the Company at the request of Parent, or (c) outside the U.S., who remain employees of the Company, or who become employees of Parent or any subsidiary of Parent, as required by applicable law.

*Director and Statutory Auditors Resignations; Appointment of Directors and Statutory Auditors; Call of the Ordinary Shareholders’ Meeting.* In the Tender Offer Agreement, the Company acknowledges that, pursuant to the Tender Offer Agreement, certain directors of the Company have delivered written resignations, respectively, to the Company and to the Purchaser, pursuant to which and subject to the Offer closing, the resignations of (a) three of such individuals will become effective as of the Acceptance Time (the “Initial Director Resignations”), and (b) up to three of the other such individuals will become effective as of immediately following the effectiveness of the Acceptance Time Board Appointments (as defined below). Furthermore, the Company will use reasonable efforts to obtain written resignations of all the Company’s statutory auditors (including the alternate members), pursuant to which each statutory auditor will resign from its office effective as of the date of the first shareholders’ meeting to be held following the Acceptance Time. The Company Board will (x) subject to the Offer closing, upon the Acceptance Time, make certain resolutions agreed to pursuant to the terms and subject to the conditions of the Tender Offer Agreement, including the appointment of certain of



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Parent's designees to the Company Board (the "Acceptance Time Board Appointments") and (y) no later than ten business days after the Acceptance Time, issue the notice of call and take all reasonable actions to cause an ordinary shareholders' meeting of the Company to be convened in compliance with the Company's bylaws, applicable laws and the Company's past practice at the offices of the Company at Piazza XX settembre 2, Villa Guardia (Como), Italy, on February 24, 2014 at 10:00 a.m. (Milan time), on first call, and on February 25, 2014 at 10:00 a.m. (Milan time), on second call, with record date on February 7, 2014, with the following agenda: (A) release of the resigning directors and (B) appointment of new members of the Company's corporate bodies to fill the vacancies created by the resigning members and any related and consequent resolutions. However, if by February 7, 2014 the closing of the Offer has not occurred, the Company Board will be free to revoke the notice of call of the shareholders' meeting. None of the parties guarantee that the resolutions set forth in this paragraph will, once voted, be valid and effective.

*Release of the Resigning Directors.* At the ordinary shareholders' meeting of the Company described in Section 12 – "Director and Statutory Auditors Resignations; Appointment of Directors and Statutory Auditors; Call of the Ordinary Shareholders' Meeting," Parent and Purchaser agree to propose, and to vote in favor of, the form resolution attached to Tender Offer Agreement.

*Obligations of Purchaser.* Parent will take all actions necessary to cause Purchaser and, following the Acceptance Time, the Company, to perform their respective obligations under the Tender Offer Agreement and to consummate the transactions contemplated by the Tender Offer Agreement including the Offer, upon the terms and subject to the conditions set forth in the Tender Offer Agreement.

*Confidentiality Agreements.* The Company will use commercially reasonable efforts to ensure that, prior to the Acceptance Time, certain identified individuals, and each other individual as reasonably requested by Parent, enter into a confidentiality and intellectual property assignment agreements in favor of the Company or its subsidiary, as applicable, in form and substance reasonably satisfactory to Parent.

*Termination Prior to the Acceptance Time.* The Tender Offer Agreement may be terminated and the Offer may be abandoned at any time prior to the Acceptance Time:

- by the mutual written consent of the Company and Parent;
- by either Parent or the Company, if the Offer has expired or been terminated in accordance with the terms of the Tender Offer Agreement without Purchaser having accepted for payment any Ordinary Shares and ADSs pursuant to the Offer on or before April 19, 2014 (the "Termination Date"), unless either Parent or the Company elects to extend this date to September 19, 2014 (the "Extended Termination Date") in the event that, on the Termination Date, the only conditions to the Offer that have not been satisfied or waived on or prior to the Termination Date are: (a) the pendency or occurrence of any action, proceeding or claim brought or threatened against the Company by any Governmental Authority and (b) either or both of (i) the Minimum Condition and (ii) the Company's certification of the accuracy of, and its compliance with, the representations, warranties and covenants made pursuant to the Tender Offer Agreement, and such conditions are likely to be satisfied by September 19, 2014 if the parties use their reasonable best efforts to do so, then either Parent or the Company may elect to extend the Termination Date, by written notice to the other prior to or on the Termination Date, until September 19, 2014 (the "Extended Termination Date").
- by either Parent or the Company, if any governmental entity of competent jurisdiction has enacted, issued, promulgated, entered, enforced or deemed applicable to the Offer any law issued or granted any order (that is final and non-appealable) that is in effect and has the effect of making the Offer illegal or that has the effect of prohibiting or otherwise preventing the Offer;



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- by the Company:
  - in the event (a) of a breach of any covenant or agreement on the part of Parent or Purchaser set forth in the Tender Offer Agreement or (b) that any of the representations and warranties of Parent or Purchaser set forth in the Tender Offer Agreement was inaccurate when made or as become inaccurate, in each case, so as to prevent Purchaser from consummating the Offer in accordance with the terms the Tender Offer Agreement), provided that in certain circumstances Parent or Purchaser may have up to 30 days to cure such breach or inaccuracy or, if earlier, until the Termination Date (or Extended Termination Date, as applicable); or
  - immediately prior to or concurrently with entering into a binding, written, definitive Acquisition Agreement providing for the consummation of a Superior Proposal (a “Specified Definitive Acquisition Agreement”) if: (a) the Company has not materially breached certain terms of the Tender Offer Agreement; (b) subject to complying with the terms of the Tender Offer Agreement, the Company Board has effected a Company Board Recommendation Change and authorized the Company to enter into the Specified Definitive Acquisition Agreement; (c) the Company, immediately prior to or concurrently with such termination, pays or causes to be paid to Parent the applicable termination fee as set forth in the Tender Offer Agreement; or
- by Parent:
  - in the event (a) of a breach of any covenant or agreement on the part of the Company set forth in the Tender Offer Agreement such that the Company has failed to perform or comply in all material respects with its obligations required to be performed or complied with by it under the Tender Offer Agreement, or (b) that any representation or warranty of the Company set forth in the Tender Offer Agreement was inaccurate when made or as becomes inaccurate, such that certain conditions to the Offer would not be satisfied, provided that in certain circumstances the Company may have up to 30 days to cure such breach or inaccuracy or, if earlier, until the Termination Date (or Extended Termination Date, as applicable); or
  - in the event that any of the following events (each, a “Triggering Event”) have occurred: (a) the Company Board or any committee thereof has for any reason effected a Company Board Recommendation Change; (b) the Company has failed to include the Company Board Recommendation in the Schedule 14D-9 or to permit Purchaser to include the Company Board Recommendation in the Offer documents; (c) the Company Board or any committee thereof has for any reason approved, or recommended that the Company’s shareholders approve, any Acquisition Proposal or Acquisition Transaction (whether or not a Superior Proposal); (d) any tender offer (including self-tender) or exchange offer that, if consummated would result in any person, entity or “group” (as defined in or under Section 13(d) of the Exchange Act) beneficially owning 15% or more of the total outstanding voting securities of the Company or any of its subsidiaries has been made by a person or entity unaffiliated with Parent or Purchaser and, within ten business days after notice of such tender offer is first published, sent or given to the Company’s shareholders (or, if earlier, prior to the Acceptance Time), the Company has not made, pursuant to Rule 14e-2 under the Exchange Act, a statement that the Company recommends rejection of such offer and unconditionally reaffirming the Company Board Recommendation; (e) an Acquisition Proposal has been publicly disclosed, announced, commenced, submitted or made, and the Company Board fails to (i) publicly and unconditionally reaffirm the Company Board Recommendation and (ii) either unconditionally reject or recommend, by the unanimous vote of all directors of the Company other than those that have deemed themselves conflicted, that the Company’s shareholders reject such Acquisition Proposal, as applicable, within ten business days after Parent requests in writing that such recommendation be reaffirmed publicly; or (f) the Company, any subsidiary of the Company or any of their respective representatives has breached any of the non-solicitation provisions described under “No Solicitation by the Company; Other Offers and such breach will have resulted in or facilitated an Acquisition Proposal being announced, submitted or made.

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The party desiring to terminate the Tender Offer Agreement must give written notice of such termination to the other party in writing specifying the provision or provisions the Tender Offer Agreement pursuant to which such termination is effected, which notice will be deemed given under the Tender Offer Agreement when delivered and received (i) immediately upon delivery personally, (ii) four business days after being sent by registered or certified mail, return receipt requested, postage prepaid, (iii) one business day after being sent for next business day delivery, fees prepaid, via a reputable international overnight commercial delivery service, or (iv) immediately if sent via facsimile (if electronic receipt is confirmed).

*Notice of Termination; Effect of Termination.* Any proper termination of the Tender Offer Agreement will be effective immediately upon the delivery of written notice by the terminating party to the other party or parties, as applicable. In the event of the termination of the Tender Offer Agreement in accordance with the terms described under “Termination Prior to the Acceptance Time,” the Tender Offer Agreement will be of no further force or effect without liability of any party or parties, as applicable (or any shareholder, director, officer, employee, agent, consultant or representative of such party or parties) to the other party or parties, as applicable, with certain exceptions, including that the payments described under “General Fees And Expenses” and “Termination Fee” will still be payable, as applicable, and that such termination will not relieve any party or parties, as applicable, from liability for any willful and material breach of, or fraud in connection with, the Tender Offer Agreement. In addition to the foregoing, no termination of the Tender Offer Agreement will affect the obligations of the parties set forth in the Confidentiality Agreement (as defined under “Other Agreements – Confidentiality Agreement”), all of which obligations will survive termination of the Tender Offer Agreement in accordance with their terms.

*General Fees and Expenses.* All fees and expenses incurred in connection with the Tender Offer Agreement and the transactions contemplated hereby (including the Offer) will be paid by the party or parties, as applicable, incurring such expenses whether or not the Offer is consummated.

### *Termination Fee.*

In the event that the Tender Offer Agreement is terminated by the Company due to the Company entering into a Specified Definitive Acquisition Agreement, the Company will pay or cause to be paid to Parent the nonrefundable amount of \$25,300,000, by wire transfer of immediately available funds to an account or accounts designated in writing by Parent, immediately prior to or concurrent with such termination.

In the event that the Tender Offer Agreement is terminated (x) by Parent for an uncured breach by the Company of its representations, warranties, or covenants contained in the Tender Offer Agreement, or (y) by Parent or the Company following the occurrence of a Triggering Event, the Company must pay to Parent, (1) by wire transfer of immediately available funds to an account or accounts designated in writing by Parent, within two business days of such termination, a non-refundable fee in the amount of \$10,100,000, and (2) if, within twelve months after the date of any such termination, a Specified Acquisition Transaction is consummated or a definitive agreement contemplating a Specified Acquisition Transaction (other than a Specified Acquisition Transaction in the form of a license of assets of the Company and/or the Company’s subsidiaries) is executed, a non-refundable fee in the amount of \$15,200,000 at the time such Specified Acquisition Transaction is consummated. A “Specified Acquisition Transaction” will mean any Acquisition Transaction; provided, however, that for purposes of this definition, all references to “15%” in the definition of Acquisition Transaction will be deemed to be references to “40%,” and all references to “85%” in the definition of Acquisition Transaction will be deemed to be references to “60%.”

In the event that the Tender Offer Agreement is terminated by Parent or the Company due to the expiration of the Offer without Purchaser having accepted Ordinary Shares or ADSs for payment and: (a) at or prior to the time of such termination an Acquisition Proposal was disclosed, announced, commenced, submitted or made; and (b) within 12 months after the date of any such termination, a Specified Acquisition Transaction (whether or not relating to such Acquisition Proposal) is consummated or a definitive agreement contemplating a Specified

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Acquisition Transaction (whether or not relating to such Acquisition Proposal) is executed, then the Company will pay (or cause to be paid) to Parent a nonrefundable amount of \$25,300,000 (such non-refundable fee, and each of the other non-refundable fees set forth in this section “Termination Fee,” a “Termination Fee”), by wire transfer of immediately available funds to an account or accounts designated in writing by Parent, at the time such Specified Acquisition Transaction is consummated. However, no termination will be payable if such Specified Acquisition Transaction is in the form of a license of assets of the Company and or the Company’s subsidiaries unless such Specified Acquisition Transaction is entered into with any person or entity that disclosed, announced, commenced, submitted or made an Acquisition Proposal prior to the date of such termination, or any of such person or entity’s affiliates.

In no event will the Company be required to pay any Termination Fee on more than one occasion, whether or not the Termination Fees may be payable under more than one of the above circumstances, at the same or at different times and upon the occurrence of different events.

Upon payment of the applicable Termination Fee(s), the Company will have no further liability or obligation to Purchaser under the Tender Offer Agreement, at law or in equity or otherwise. Each of the parties acknowledge that a Termination Fee is not a penalty, but rather is liquidated damages in a reasonable amount that will compensate the Purchaser in the circumstances in which such Termination Fee is payable. However, the foregoing two sentences do not apply to any breach of the Tender Offer Agreement by the Company, including any breach that results in the termination by Parent for a breach of the Tender Offer Agreement by the Company, or in the event of fraud.

*Amendment.* Subject to applicable law and subject to the other provisions of the Tender Offer Agreement, the Tender Offer Agreement may be amended by the parties at any time by execution of an instrument in writing signed on behalf of each of Parent and the Company. To the extent that any amendment or modification to certain terms of the Tender Offer Agreement are sought which are adverse to the rights of any debt financing sources, the prior written consent of such debt financing sources party to the debt financing will be required before such amendment or modification is rendered effective.

*Extension; Waiver.* At any time and from time to time prior to the Acceptance Time, any party or parties hereto may, to the extent legally allowed and except as otherwise set forth in the Tender Offer Agreement, (a) extend the time for the performance of any of the obligations or other acts of the other party or parties, as applicable, (b) waive any inaccuracies in the representations and warranties made to such party or parties contained in the Tender Offer Agreement or in any document delivered pursuant hereto and (c) waive compliance with any of the agreements or conditions for the benefit of such party or parties contained in the Tender Offer Agreement. Any agreement on the part of a party or parties to any such extension or waiver will be valid only if set forth in an instrument in writing signed on behalf of such party or parties, as applicable. Any delay in exercising any right under the Tender Offer Agreement will not constitute a waiver of such right.

## **Other Agreements**

*Support Agreements.* In connection with the Tender Offer Agreement, each of Khalid Islam, Chief Executive Officer of the Company, Giorgio Mosconi, Vice President, Medical Affairs of the Company, Adrian Haigh, Senior Vice President, Commercial Operations of the Company, Sal Calabrese, Chief Financial Officer of the Company, and Carin Heringa, Scientific Director and Senior Vice President of the Company, each member of the Company Board (other than Marco Brughera), and F3F S.r.l., the beneficial owner of approximately [15%] of the outstanding Ordinary Shares and ADSs that is represented on the Company Board, entered into support agreements with Parent (collectively, the “Support Agreements”), pursuant to which they each agreed, among other things, to:

- promptly tender all of their Ordinary Shares and ADSs into the Offer;
- not transfer their Ordinary Shares and ADSs, subject to certain limited exceptions;

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- exercise any options to acquire ADSs held by them following the Acceptance Time and tender the underlying ADSs into the Offer during the subsequent offering period;
- vote their Ordinary Shares and ADSs against the following actions (and, except in the case of F3F S.r.l., deliver a proxy or proxies to Parent if any Company shareholders' meeting is called during the term of the Support Agreement): (i) any Acquisition Transaction; (ii) any change in a majority of the board of directors of the Company; (iii) any amendment to the Company's articles of association or bylaws; (iv) any material change in the capitalization of the Company or the Company's corporate structure; and (v) any other action which is intended, or could reasonably be expected, to impede, interfere with, delay, postpone, discourage or adversely affect the Offer or any of the other transactions contemplated by the Tender Offer Agreement or the Support Agreement;
- not (i) solicit, initiate or knowingly encourage (including by way of providing non-public information) the submission or announcement of any inquiries, proposals or offers that constitute or would reasonably be expected to result in an Acquisition Proposal; (ii) provide any non-public information concerning the Company or any of its subsidiaries to any individual or entity in connection with or in response to an Acquisition Proposal; (iii) engage in any discussions or negotiations with any individual or entity with respect to any Acquisition Proposal; (iv) approve, adopt, endorse or recommend any Acquisition Proposal; (v) enter into any letter of intent or similar document or any Contract contemplating or otherwise relating to any Acquisition Transaction; or (vi) continue any existing discussions with any individual or entity that relate to any Acquisition Proposal; and
- for a period of 18 months following the Acceptance Time, not, directly or indirectly, encourage, induce, attempt to induce, solicit or attempt to solicit (on their own behalf or on behalf of any other individual or entity) any current or former employee of the Company to leave his or her employment, consulting or independent contractor relationship with the Company or any of the Company's subsidiaries (subject to customary exceptions).

Each of the Support Agreements also provides that, if the Tender Offer Agreement is terminated (a) by the Company in order to accept a superior proposal; (b) following the occurrence of a Triggering Event or (c) as a result of the expiration of the Offer without Purchaser having accepted any Ordinary Shares or ADSs for payment and an Acquisition Proposal was disclosed, announced, commenced, submitted or made, at or prior to the time of such termination, then the restrictions on transfer of Ordinary Shares and ADSs and the voting covenants contained therein will continue in effect for a period of six months following the termination of the Tender Offer Agreement.

Approximately 4,016,608 Ordinary Shares and ADSs (including 1,666,608 ADSs issuable upon the exercise of options) are subject to Support Agreements (representing approximately 15% of the total outstanding Ordinary Shares and ADS, and approximately 23% of the fully diluted number of Ordinary Shares and ADSs).

In addition, each of the Support Agreements, other than the Support Agreements with F3F S.r.l. and Carin Heringa, contain a provision prohibiting the signatory, for a period of three years following the Acceptance Time, from: (i) having directly or indirectly, any financial or other interest in a business or company which engages in the field of: manufacture, production, promotion, sale and commercialization of (A) any agent for the treatment or prevention of veno-occlusive disease (sinusoidal obstruction syndrome) or graft versus host disease, and (B) any polydisperse agent comprising deoxyribonucleic acid fragments (a "Competitor"); (ii) accepting any part or full time employment in a Competitor or to act as a consultant or representative or in any other form for a Competitor; (iii) directly or indirectly establishing a Competitor; (iv) enticing away or solicit or try to entice away or to solicit any customer of the Company or any of the Company's subsidiaries for whom each was active or with whom each was in contact during the last 12 months prior to the Acceptance Time (a "Customer"); or (v) directly nor indirectly acting on behalf of or rendering services to a Customer, for example as an employee, consultant, agent, officer, director nor submit or be involved in any offer for such activity; provided however, that Mr. Khalid is permitted to perform certain consulting services.

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*Transition, Amendment and Release Agreement.* Concurrently with the execution of the Tender Offer Agreement, Dr. Islam entered into a transition, amendment and release agreement with the Company (the “Transition, Amendment and Release Agreement”), whereby he has agreed to resign from the Company on the Closing Date (as defined in his Transition, Amendment and Release Agreement), and that he will only be eligible to receive certain change in control severance entitlements provided that he successfully completes his Consultancy Agreement.

The foregoing description of the Transition, Amendment and Release Agreement does not purport to be complete and is qualified in its entirety by reference to the Transition, Amendment and Release Agreement, a form of which is filed as Exhibit (d)(3) to the Schedule TO, and is incorporated herein by reference.

*Consultancy Agreement.* Concurrently with the execution of the Tender Offer Agreement, Dr. Islam entered into a consultancy agreement with the Company (the “Consultancy Agreement”), whereby he has agreed to work as a consultant for the Company for the six months following the Closing Date, as defined in the Transition, Amendment and Release Agreement.

The foregoing description of the Consultancy Agreement does not purport to be complete and is qualified in its entirety by reference to the Consultancy Agreement, a form of which is filed as Exhibit (d)(4) to the Schedule TO, and is incorporated herein by reference.

*Retention and Amendment Agreements.* Concurrently with the execution of the Tender Offer Agreement, each of Mr. Calabrese, Dr. Mosconi, and Mr. Haigh entered into a Retention and Amendment Agreement (collectively, the “Retention Agreements”), whereby each individual has agreed to continue working for the Company or a wholly owned subsidiary of the Company for a period of six to nine months, respectively, as applicable, and receive a retention bonus for this period. Further, each agreed that he will not be entitled to receive certain change in control severance entitlements (as defined in the respective employment agreements), or in the case of Mr. Haigh and Mr. Calabrese, he resigns without Good Reason, as defined in the respective employment agreements, prior to the completion of the Retention Period, as defined in the respective Retention Agreements.

The foregoing description of the Retention Agreements does not purport to be complete and is qualified in its entirety by reference to the Retention Agreements, a form of which is filed as Exhibit (d)(5) to the Schedule TO, and is incorporated herein by reference.

*Confidentiality Agreement.* Parent and the Company entered into a Confidentiality and Nondisclosure Agreement regarding non-disclosure and other obligations, effective as of September 30, 2013 (the “Confidentiality Agreement”), during the course of discussions regarding a potential acquisition of the Company. Under the Confidentiality Agreement, Parent agreed, subject to certain exceptions, to keep non-public information concerning the Company confidential.

The foregoing description of the Confidentiality Agreement does not purport to be complete and is qualified in its entirety by reference to the Confidentiality Agreement, which is filed as Exhibit (d)(6) to the Schedule TO, and is incorporated herein by reference.

### **13. Purpose of the Offer; Plans for the Company.**

*Purpose of the Offer.* The purpose of the Offer is for Parent, through Purchaser, to acquire control of, and ultimately the entire equity interest in, the Company. The Offer is intended to facilitate the acquisition of all outstanding Ordinary Shares and ADSs, or, at a minimum, permit Parent to control a majority interest in the Company.

The Company Board has approved the Tender Offer Agreement and the transactions contemplated thereby, including the Offer.

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*Plans for the Company.* Except as otherwise provided in the Tender Offer Agreement, it is expected that, initially following the Acceptance Time, the business and operations of the Company will be continued substantially as they are currently being conducted. Parent will continue to evaluate the business and operations of the Company over time and will take such actions with respect to the Company as it deems appropriate under the circumstances then existing. From time to time, Parent may make changes to the Company business, operations, capitalization or management with a view to combining the existing and future business of the Company and Parent and optimizing the Company's business in conjunction with Parent's other business.

Except as set forth in this Offer to Purchase, Parent and Purchaser have no present plans or proposals that would relate to or result in (i) any extraordinary corporate transaction involving the Company or any of its subsidiaries (such as a merger, reorganization, liquidation, relocation of any operations or sale or other transfer of a material amount of assets), (ii) any sale or transfer of a material amount of assets of the Company or any of its subsidiaries, (iii) any material change in the Company capitalization or dividend policy or (iv) any other material change in the Company corporate structure or business.

### **14. Certain Conditions of the Offer.**

For the purposes of this Section 14, capitalized terms used but not defined herein will have the meanings set forth in the Tender Offer Agreement. Notwithstanding any other provisions of the Offer, in addition to (and not in limitation of) the rights and obligations of Purchaser to extend and/or amend the Offer at any time in its sole discretion (subject to the terms and conditions of the Tender Offer Agreement), Purchaser (i) will not be required to accept for payment or, subject to any applicable rules and regulations of the SEC (including Rule 14e-1(c) under the Exchange Act (relating to the obligation of Purchaser to pay for or return tendered Ordinary Shares and ADSs promptly after termination or withdrawal of the Offer)), pay for, and (ii) may delay the acceptance for payment of or, subject to the restriction referred to above, the payment for, any tendered Ordinary Shares and ADSs, in the event that at or prior to the scheduled Offer Closing (as it may be extended pursuant to terms of the Tender Offer Agreement) if:

(A) the Minimum Condition (as it may have been modified by Purchaser electing to effect a Permitted Minimum Condition Modification) has not been satisfied,

(B) any of the following has occurred and is continuing to occur as of the scheduled expiration of the Offer:

(1) any of the representations listed below ("Specified Representations") were not true and correct in all material respects as of the date of the Tender Offer Agreement or are not true and correct in all material respects as of the scheduled expiration of the Offer with the same force and effect as if made as of the scheduled expiration of the Offer (except to the extent expressly made as of an earlier date, in which case as of such date):

- capital stock;
- authorization;
- applicability of takeover statutes;
- no vote being required;
- opinion of financial advisor; or
- the Company's compliance with Rule 14d-10 under the Exchange Act.

provided, however, that, for purposes of determining whether such representations and warranties are true and correct any update of or modification to the Company Disclosure Letter made or purported to have been made after the execution and delivery of the Tender Offer Agreement will be disregarded;

(2) any of the representations and warranties of the Company set forth in the Tender Offer Agreement, other than the Specified Representations, were not true and correct as of the date of the Tender Offer

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Agreement or are not true and correct as of the scheduled expiration of the Offer with the same force and effect as if made as of the scheduled expiration of the Offer (except to the extent expressly made as of an earlier date, in which case as of such date), except that any failure of any of such representations and warranties to be true and correct will be disregarded if the circumstances giving rise to such failures to be true and correct would not, individually or in the aggregate, reasonably be expected to have or result in a Company Material Adverse Effect; provided, however, that, for purposes of determining whether such representations and warranties are true and correct: (x) all materiality or “Company Material Adverse Effect” qualifications limiting the scope of such representations and warranties will be disregarded; and (y) any update of or modification to the Company Disclosure Letter made or purported to have been made after the execution and delivery of the Agreement will be disregarded;

(3) the Company fails to perform or comply in all material respects with its obligations required to be performed or complied with by it under the Agreement;

(4) since the date of the Agreement, any Company Material Adverse Effect shall have occurred;

(5) Parent does not receive a certificate executed by the Company’s Chief Executive Officer and Chief Financial Officer confirming that the conditions set forth in clauses “(B)(1),” “(B)(2)” and “(B)(3)” of this Section 14 – “Certain Conditions of the Offer” have been duly satisfied as of the scheduled expiration of the Offer;

(6) any temporary restraining order, preliminary or permanent injunction or other Order preventing the acquisition of or payment for Ordinary Shares and ADSs pursuant to the Offer has been issued by any court of competent jurisdiction or other Governmental Entity and remains in effect, or any law is enacted or deemed applicable to the Offer that makes the acquisition of or payment for Ordinary Shares and ADSs pursuant to the Offer illegal or violative of any applicable law;

(7) there is any pending or overtly threatened action, claim or proceeding by any governmental entity of competent jurisdiction against Parent, Purchaser, the Company or any Subsidiary of the Company: (x) challenging or seeking to restrain or prohibit the acquisition of or payment for Ordinary Shares and ADSs pursuant to the Offer or any of the other transactions contemplated by the Tender Offer Agreement; (y) seeking to prohibit or limit in any material respect Parent or Purchaser’s ability to vote, transfer, receive dividends with respect to or otherwise exercise ownership rights with respect to the stock of the Company; or (z) seeking to compel Parent, Purchaser, the Company or any of their respective Subsidiaries to dispose of or hold separate any Ordinary Shares and ADSs or any material assets as a result of the Offer or any of the other transactions contemplated by the Tender Offer Agreement;

(8) Parent does not receive a resolution approved by the Company Board in the form set forth in Schedule 2 to the Tender Offer Agreement, pursuant to which, among other things, the Company Board appoints three nominees of Parent to fill vacancies on the Company Board; or

(9) the Agreement is terminated in accordance with its terms.

The foregoing conditions (other than the Minimum Condition, as it may be modified by Purchaser electing to effect a Permitted Minimum Condition Modification) are for the sole benefit of Parent and Purchaser and, subject to the terms and conditions of the Agreement, may be waived by Parent or Purchaser, in whole or in part at any time and from time to time in the sole discretion of Parent and Purchaser. The failure by Parent or Purchaser at any time to exercise any of the foregoing rights shall not be deemed a waiver of any such right and each such right shall be deemed an ongoing right which may be asserted at any time and from time to time.

As used in the Tender Offer Agreement, a “Company Material Adverse Effect” is any fact, circumstance, occurrence, event, change or effect (an “Effect”) that, individually or when taken together with all other Effects,

has had or is reasonably likely to have a material adverse effect on the business, operations, assets (including intangible assets), financial condition or results of operations of the Company taken as a whole with its Subsidiaries, provided, however, that, none of the following Effects, by itself or when aggregated with any one or more other Effects, will be deemed to be or constitute a Company Material Adverse Effect and none of the following Effects, by itself or when aggregated with any one or more other effects, will be taken into account when determining whether a Company Material Adverse Effect has occurred or is reasonably likely to occur: effects arising out of or resulting from (a) (ii) general market, economic or political conditions in Italy or worldwide or (ii) conditions (or any changes therein) in the industries in which the Company or any of its subsidiaries conduct business, in each case, including any acts of terrorism or war, weather conditions or other force majeure events, in the case of each of clauses (i) and (ii), to the extent that such effects do not have and are not reasonably likely to have a disproportionate impact on the Company and its subsidiaries, taken as a whole, relative to other companies operating in the same industries in which the Company or any of its subsidiaries conduct business; (b) the announcement of the execution of the Tender Offer Agreement or the pendency of the Offer, including any loss of, or adverse change in, the relationship of the Company or any of its subsidiaries with its employees, licensors, licensees, customers, distributors, partners or suppliers resulting or arising therefrom but excluding any claim or action made or brought by any holder of Ordinary Shares or ADSs in its capacity as a shareholder which, together with its affiliates, beneficially owns 10% or more of the aggregate number of Ordinary Shares or ADSs (without duplication for Ordinary Shares underlying the ADSs); (c) changes in accounting standards (or the interpretation thereof by a third party); (d) changes in applicable law or regulatory conditions (or the interpretation thereof by a third party), to the extent that such changes do not have a disproportionate impact on the Company and its subsidiaries, taken as a whole, relative to other companies operating in the same industries in which the Company or any of its subsidiaries conduct business; (e) changes in the trading price or trading volume of the ADSs, in and of themselves (it being understood that any underlying cause of any such change may, subject to the other terms of this definition, be taken into consideration when determining whether a Company Material Adverse Effect has occurred or is reasonably likely to occur); or (f) any failure by the Company or any of its subsidiaries to meet any public estimates or expectations of the Company's bookings, revenue, earnings or other financial performance or results of operations for any period, or any failure by the Company or any of its subsidiaries to meet any internal budgets, plans or forecasts of its bookings, revenues, earnings, earnings per share or other financial performance or results of operations (it being understood that any underlying cause of any such failure may, subject to the other terms of this definition, be taken into consideration when determining whether a Company Material Adverse Effect has occurred or is reasonably likely to occur).

#### **15. Certain Legal Matters; Regulatory Approvals.**

*General.* Except as described in this Section 15, the Company has represented to us in the Tender Offer Agreement that no consent, approval, order or authorization of, or registration, declaration, filing with or notice to any Governmental Entity (as defined below) is required by or with respect to the Company or any of its Subsidiaries in connection with the execution, delivery and performance of the Tender Offer Agreement by the Company or the consummation of the Offer or compliance with the provisions of the Tender Offer Agreement, except for such filings and reports as required by the applicable requirements of the antitrust laws, the Securities Act, the Exchange Act, the rules and regulations promulgated thereunder, and state securities laws or "blue sky" laws, any filings required under the rules and regulations of Nasdaq and such other filings the failure of which to be obtained or made, individually or in the aggregate, have not had and would not be reasonably expected to have a Company Material Adverse Effect or would not prevent, materially delay or materially impede consummation of the Offer. "Governmental Entity" means any EU, EU member state, United States, federal, state, provincial, local, municipal, district, foreign or other jurisdiction of any nature government, any government or subdivision thereof or any other governmental, administrative, judicial, arbitral, legislative, executive, regulatory or self-regulatory authority, instrumentality, agency, commission or body. Should any such approval or other action be required, we currently contemplate that, except as described below under the paragraph relating to takeover laws below, such approval or other action will be sought. While we do not currently intend to delay acceptance for payment of Ordinary Shares and ADSs tendered pursuant to the Offer pending the outcome of any such matter,



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there can be no assurance that any such approval or other action, if needed, would be obtained or would be obtained without substantial conditions. If such approvals were not obtained or such other actions were not taken, adverse consequences might result to the Company's business, of which under certain conditions specified in the Tender Offer Agreement, could permit us to elect to terminate the Offer without the purchase of Ordinary Shares and ADSs thereunder under. See Section 14 – "Certain Conditions of the Offer."

*Takeover Laws.* The Company is incorporated under the laws of the Republic of Italy. The Company represented to us in the Tender Offer Agreement that to the knowledge of the Company, the Company Board has taken all actions necessary to ensure that no "moratorium," "fair price," "business combination," "control share acquisition" or similar provision of any state anti-takeover law is, or at the Offer closing will be, applicable to the Tender Offer Agreement or the Support Agreements.

*Legal Proceedings.* The Company represented to us in the Tender Offer Agreement that there is no action, claim, suit, litigation, proceeding (public or private) or criminal prosecution by or before any Governmental Entity pending or, to the knowledge of the Company, threatened in writing seeking to impose any legal restraint on or prohibition against the Offer or that, if resolved adversely to the Company, would have the effect of preventing or materially delaying, or making illegal, the Offer.

*Exchange Act Registration.* The Ordinary Shares and ADSs are currently registered under the Exchange Act. The purchase of the Ordinary Shares and ADSs pursuant to the Offer may result in the Ordinary Shares and ADSs becoming eligible for deregistration under the Exchange Act. Registration of the Ordinary Shares and ADSs may be terminated by the Company upon application to the SEC if the outstanding Ordinary Shares and ADSs are not listed on a "national securities exchange" and there are fewer than 300 holders of record of Ordinary Shares and ADSs.

Termination of registration of the Ordinary Shares and ADSs under the Exchange Act would reduce the information required to be furnished by the Company to holders of its securities and to the SEC and would make certain provisions of the Exchange Act, such as the requirement of furnishing annual reports to security holders, no longer applicable with respect to the Company. Furthermore, the ability of "affiliates" of the Company and persons holding "restricted securities" of the Company to dispose of such securities pursuant to Rule 144 under the Securities Act of 1933, as amended, may be impaired or eliminated. If registration of the Ordinary Shares and ADSs under the Exchange Act were terminated, the Ordinary Shares and ADSs would no longer be eligible for continued inclusion on the Federal Reserve Board's list of "margin securities" or eligible for stock exchange listing or reporting on Nasdaq.

### **16. Appraisal Rights.**

Under Italian law, the holders of Ordinary Shares and ADSs who do not tender their Ordinary Shares or ADSs in the Offer will not have dissenters rights or appraisal rights in connection with the Offer. It is also doubtful whether under Italian law, upon delisting, holders of Ordinary Shares and ADSs may be entitled to exercise withdrawal rights with respect to such securities. Should a withdrawal right apply, the cash amount to which holders exercising withdrawal rights would be entitled in the event of a delisting would be equal to the arithmetic average of the closing prices of the ADSs during the six months preceding publication of the call notice convening the Company's shareholders' meeting approving the delisting.

### **17. Fees and Expenses.**

Neither Purchaser nor Parent will pay any fees or commissions to any broker or dealer or other person for soliciting tenders of Ordinary Shares and ADSs pursuant to the Offer. Brokers, dealers, commercial banks and trust companies will, upon request, be reimbursed by Purchaser for reasonable and necessary mailing and handling expenses incurred by them in forwarding material to their customers.

The Purchaser has retained MacKenzie Partners, Inc. as Information Agent, and The Bank of New York Mellon as Tender Agent in connection with the Offer. The Information Agent and the Tender Agent will receive

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reasonable and customary compensation for their services hereunder and reimbursement for their reasonable out-of pocket expenses. The Information Agent and the Tender Agent also will be indemnified by Purchaser against certain liabilities in connection with the Offer.

### **18. Miscellaneous.**

The Offer is not being made to (nor will tenders be accepted from or on behalf of) holders of Ordinary Shares and ADSs in any jurisdiction in which the making of the Offer or the acceptance thereof would not be in compliance with the securities, blue sky or other laws of such jurisdiction. In those jurisdictions where applicable laws require the Offer to be made by a licensed broker or dealer, the Offer shall be deemed to be made on behalf of Purchaser by one or more registered brokers or dealers licensed under the laws of such jurisdiction to be designated by Purchaser.

**No person has been authorized to give any information or make any representation on behalf of Purchaser other than as contained in this Offer to Purchase or in the Share Form of Acceptance or ADS Letter of Transmittal, and, if any such information or representation is given or made, it should not be relied upon as having been authorized by Purchaser.**

The Purchaser has filed with the SEC a Tender Offer Statement on Schedule TO pursuant to Rule 14d-3 under the Exchange Act, together with exhibits furnishing certain additional information with respect to the Offer, and may file amendments thereto. In addition, the Company will file with the SEC a Schedule 14D-9, together with exhibits, pursuant to Rule 14d-9 under the Exchange Act, setting forth the recommendation of the Company's Board with respect to the Offer and the reasons for such recommendation and furnishing certain additional related information. A copy of such documents, and any amendments thereto, may be examined at, and copies may be obtained from, the SEC in the manner set forth under Section 8 – "Certain Information Concerning the Company" above.

Jazz Pharmaceuticals Italy S.r.l.

December 23, 2013

**SCHEDULE I  
DIRECTORS AND EXECUTIVE OFFICERS OF THE PURCHASER AND PARENT**

**1. PURCHASER**

The name, business address, present principal occupation or employment and material occupations, positions, offices or employment for the past five years of the sole director of Purchaser are set forth below. The business address of Purchaser is c/o Jazz Pharmaceuticals plc, Fourth Floor, Connaught House, One Burlington Road, Dublin 4, Ireland. The telephone number at such office is +011-353-1-634-7800.

**NAME AND POSITION**  
Fintan Keegan, Director

**PRESENT PRINCIPAL OCCUPATION OR EMPLOYMENT**

Mr. Keegan was appointed Executive Vice President, Technical Operations of Parent in July 2012 and served as its Senior Vice President of Technical Operations from the Azur Merger until July 2012. Prior to the Azur Merger, he was Senior Vice President and Chief Technical Officer of Azur Pharma from 2006 until the Azur Merger, where he was responsible for quality, regulatory, compliance, supply chain and development. Prior to his work with Azur Pharma, Mr. Keegan most recently served as Vice President of Quality and Regulatory for Elan Corporation, plc. He also held various positions with Wyeth Pharmaceuticals, Inc., Merck & Co., Inc. and at a clinical contract research organization. Mr. Keegan holds a B.Sc and a H. Dip in Pharmaceutical Manufacturing Technology from Trinity College Dublin and a M.Sc from the School of Chemistry, University of Bristol, in the United Kingdom. Mr. Keegan is a citizen of Ireland.

**2. PARENT**

The name, business address, present principal occupation or employment and material occupations, positions, offices or employment for the past five years of each of the directors and executive officers of Parent are set forth below. The business address and phone number of each such director and executive officer is c/o Jazz Pharmaceuticals plc, Fourth Floor, Connaught House, One Burlington Road, Dublin 4, Ireland. The telephone number at such office is +011-353-1-634-7800. Unless otherwise indicated, each director and executive officer is a citizen of the United States of America.

**NAME AND POSITION**  
Paul L. Berns, Director

**PRESENT PRINCIPAL OCCUPATION OR  
EMPLOYMENT AND EMPLOYMENT HISTORY**

Mr. Berns has served as a member of Parent's board of directors since the Azur Merger and was a director of Jazz Pharmaceuticals, Inc. from 2010 until the Azur Merger. Mr. Berns is a self-employed consultant to the pharmaceutical industry. From March 2006 to September 2012, he served as the President and Chief Executive Officer, and as a member of the board of directors, of Allos Therapeutics, Inc., a pharmaceutical company acquired by Spectrum Pharmaceuticals, Inc. From July 2005 to March 2006, Mr. Berns was a self-employed consultant to the pharmaceutical industry. From June 2002 to July 2005, Mr. Berns was President, Chief Executive Officer and a director of Bone Care International, Inc., a specialty pharmaceutical company that was acquired by Genzyme Corporation in 2005. From 2001 to 2002, Mr. Berns served as Vice President and General Manager of the Immunology, Oncology and Pain Therapeutics business unit of Abbott Laboratories, a pharmaceutical company. From 2000 to 2001, he served as Vice President, Marketing of BASF Pharmaceuticals/Knoll, a pharmaceutical company, and from 1990 to 2000, Mr. Berns held

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**NAME AND POSITION**

**PRESENT PRINCIPAL OCCUPATION OR  
EMPLOYMENT AND EMPLOYMENT HISTORY**

various positions, including senior management roles, at Bristol-Myers Squibb Company, a pharmaceutical company. Mr. Berns is chairman of the board of directors of Anacor Pharmaceuticals, Inc., which board he joined in June 2012, and has been a director of XenoPort, Inc. since 2005. Mr. Berns received a B.S. in Economics from the University of Wisconsin.

Bruce C. Cozadd, Director, Chairman of the Board and Chief Executive Officer

Mr. Cozadd has served as Parent's Chairman and Chief Executive Officer since the Azur Merger. He was a co-founder and has served (and continues to serve) as Chairman and Chief Executive Officer of Jazz Pharmaceuticals, Inc. since April 2009. From 2003 until 2009, he served as Jazz Pharmaceuticals, Inc.'s Executive Chairman and as a member of its board of directors. From 1991 until 2001, he held various positions with ALZA Corporation, a pharmaceutical company acquired by Johnson & Johnson, most recently as its Executive Vice President and Chief Operating Officer, with responsibility for research and development, manufacturing and sales and marketing. Previously at ALZA Corporation he held the roles of Chief Financial Officer and Vice President, Corporate Planning and Analysis. He serves on the boards of Cerus Corporation, a biomedical products company, Threshold Pharmaceuticals, a clinical stage biopharmaceutical company, and The Nueva School and Stanford Hospital and Clinics, both non-profit organizations. He received a B.S. from Yale University and an M.B.A. from the Stanford Graduate School of Business.

Patrick G. Enright, Director

Mr. Enright has served as a member of Parent's board of directors since the Azur Merger and was a director of Jazz Pharmaceuticals, Inc. from 2009 until the Azur Merger. Since 2006, Mr. Enright has served as a Managing Director of Longitude Capital, a venture capital firm, of which he is a founder. From 2002 through 2006, Mr. Enright was a Managing Director of Pequot Ventures, a venture capital investment firm, where he co-led the life sciences investment practice. Mr. Enright also has significant life sciences operations experience, beginning his career more than 25 years ago at Sandoz (now Novartis), a pharmaceutical company. He currently serves on the boards of directors of Corcept Therapeutics Incorporated, a pharmaceutical company, and several privately held companies. In the past five years he also served as a director of Threshold Pharmaceuticals Inc. and Sequenom Inc. Mr. Enright received a B.S. from Stanford University and an M.B.A. from the Wharton School at the University of Pennsylvania.

Peter Gray, Director

Mr. Gray has served as a member of Parent's board of directors since May 2013. Mr. Gray currently serves as Chairman of the board of directors of United Drug plc, an international provider of healthcare services, and as a business consultant to the pharmaceutical industry. In 2011, Mr. Gray retired from his position as the Chief Executive Officer of ICON plc, a global provider of outsourced development services to the pharmaceutical, biotechnology and medical device industries, which he held since November 2002. At ICON plc, Mr. Gray previously served as Group Chief Operating Officer from June 2001 to November 2002 and Chief Financial Officer from June 1997 to June 2001. Mr. Gray

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NAME AND POSITION	PRESENT PRINCIPAL OCCUPATION OR EMPLOYMENT AND EMPLOYMENT HISTORY
Heather Ann McSharry, Director	<p>holds a degree in law from Trinity College Dublin and is a chartered accountant. Mr. Gray is a citizen of Ireland.</p> <p>Ms. McSharry has served as a member of Parent’s board of directors since May 2013. Ms. McSharry currently serves as a non-executive director on the boards of directors of several public and private companies, including Greencore Group plc, an international manufacturer of convenience foods, and CRH plc, an international building materials group. From 2006 to 2009, Ms. McSharry was Managing Director Ireland of Reckitt Benckiser, a multinational health, home and hygiene consumer products company. From 1989 to 2006, she held various positions at Boots Healthcare, a leading global consumer healthcare company, most recently as Managing Director of Boots Healthcare Ireland Limited. Ms. McSharry also serves on the board of directors of the Industrial Development Agency in Ireland, where she is Chair of the audit and finance committee. From 2007 to 2011, Ms. McSharry served on the board of directors of the Bank of Ireland, including serving on its audit committee from 2009 to 2011. Ms. McSharry holds a Bachelor of Commerce and a Master of Business Studies degree from University College Dublin. Ms. McSharry is a citizen of Ireland.</p>
James C. Momtazee, Director	<p>Mr. Momtazee has served as a member of Parent’s board of directors since the Azur Merger and was a director of Jazz Pharmaceuticals, Inc. from 2004 until the Azur Merger. He is a member of KKR Management LLC, the general partner of KKR &amp; Co. L.P., and he has been employed by Kohlberg Kravis Roberts &amp; Co. L.P., or KKR, a private equity investment firm, since 1996. Funds affiliated with KKR are collectively one of Parent’s largest shareholders. He serves on the boards of directors of HCA Inc., a healthcare services company, and Accellent Inc., a manufacturing and engineering services company. He received an A.B. from Stanford University and an M.B.A. from the Stanford Graduate School of Business.</p>
Seamus Mulligan, Director	<p>Mr. Mulligan has served as a member of Parent’s board of directors since the Azur Merger and was a founder and principal investor of Azur Pharma. Mr. Mulligan serves as the Chief Executive Officer and Chairman of Adapt Pharma LLC, a specialty pharmaceutical company he founded in 2013. Since 2006, Mr. Mulligan has served as the Executive Chairman of Circ Pharma Limited and its subsidiaries, a pharmaceutical development stage group. Mr. Mulligan served as Parent’s Chief Business Officer, International Business Development from the Azur Merger until February 2013. Mr. Mulligan served as Azur Pharma’s Chairman and Chief Executive Officer and as a member of its board of directors from 2005 until the Azur Merger. From 1984 until 2004, he held various positions with Elan Corporation, a pharmaceutical company, most recently as its Executive Vice President, Business and Corporate Development. Previously at Elan Corporation, he held the roles of President of Elan Pharmaceutical Technologies, the drug delivery division of Elan, Executive Vice President, Pharmaceutical Operations, Vice President, U.S. Operations and Vice President, Product</p>

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NAME AND POSITION	PRESENT PRINCIPAL OCCUPATION OR EMPLOYMENT AND EMPLOYMENT HISTORY
	Development. He served as a member of the board of directors of the U.S. National Pharmaceutical Council until 2004. Mr. Mulligan received a B.Sc (Pharm) and M.Sc from Trinity College, Dublin, Ireland. Mr. Mulligan is a citizen of Ireland.
Kenneth W. O’Keefe, Director	Mr. O’Keefe has served as a member of Parent’s board of directors since the Azur Merger and was a director of Jazz Pharmaceuticals, Inc. from 2004 until the Azur Merger. Since January 2011 he has been Managing Partner of, and from 1997 to January 2011, he was Managing Director of, Beecken Petty O’Keefe & Company, a private equity firm, which he co-founded. He serves on the boards of several privately held healthcare companies. He received a B.A. from Northwestern University and an M.B.A. from the University of Chicago.
Norbert G. Riedel, Ph.D, Director	Dr. Riedel has served as a member of Parent’s board of directors since May 2013. Dr. Riedel is a retired executive of the pharmaceutical industry. From 2001 to January 2013, he served as Corporate Vice President and Chief Scientific Officer of Baxter International Inc., a diversified healthcare company, where from 1998 to 2001, he also served as President and General Manager of the recombinant therapeutic proteins business unit and Vice President of Research and Development of the bioscience business unit. From 1996 to 1998, Dr. Riedel served as head of worldwide biotechnology and worldwide core research functions at Hoechst-Marion Roussel, now Sanofi-Aventis, a global pharmaceutical company. Dr. Riedel serves on the board of directors of Ariad Pharmaceuticals, Inc., a biotechnology company focused on cancer, and the board of directors of the Illinois Biotechnology Industry Organization. Dr. Riedel is also a member of the Austrian Academy of Sciences, the advisory board of Northwestern University’s Kellogg School of Management Center for Biotechnology, and the Illinois Innovation Council. Dr. Riedel is an Adjunct Professor at Boston University School of Medicine and an Adjunct Professor of Medicine at Northwestern University’s Feinberg School of Medicine. Dr. Riedel holds a Diploma in biochemistry from the University of Frankfurt and a Ph.D. in biochemistry from the University of Frankfurt. Dr. Riedel is a citizen of Germany.
Catherine A. Sohn, Pharm.D., Director	Dr. Sohn has served as a member of Parent’s board of directors since her election at the July 2012 annual general meeting of shareholders. Dr. Sohn is the founder of Sohn Health Strategies, where since 2010 she has consulted to pharmaceutical, biotechnology, medical device, and consumer healthcare companies in the areas of business strategy, business development and strategic product development. She has served as a director of Landec Corporation, a material sciences company, since November 2012. From 1982 to 2010, she was with GlaxoSmithKline plc, a pharmaceutical company (and with SmithKline Beecham plc before its merger with GlaxoWellcome plc), where she served most recently as Senior Vice President, Worldwide Business Development and Strategic

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Alliances in the GSK Consumer Healthcare division, and before that, she held a series of positions in Medical Affairs, Pharmaceutical Business Development, U.S. Product Marketing, and global strategic product development in the pharmaceutical division. Dr. Sohn started her career as Assistant Professor of Clinical Pharmacy at the University of the Sciences in Philadelphia, where she currently holds the position of Dean's Professor. She received a Pharm.D. from the University of California, San Francisco, School of Pharmacy. She also received a Certificate of Professional Development from The Wharton School at the University of Pennsylvania.

Rick E. Winningham, Director

Mr. Winningham has served as a member of Parent's board of directors since the Azur Merger and was a director of Jazz Pharmaceuticals, Inc. from 2010 until the Azur Merger. Since 2001, he has served as the Chief Executive Officer and since 2010, as Chairman of the board of directors of Theravance, Inc., a biopharmaceutical company. From 1986 to 2001, he held various positions with a pharmaceutical company Bristol-Myers Squibb and its predecessor Bristol-Myers, including serving as the President of Bristol-Myers Squibb Oncology/Immunology/Oncology Therapeutics Network and, from 2000 to 2001, as its President of Global Marketing. He is a member of the board of directors of the California Healthcare Institute (CHI) and is also a member of the external advisory board of directors for the College of Business and Administration and Business Hall of Fame at Southern Illinois University. Mr. Winningham holds an M.B.A. from Texas Christian University and a B.S. from Southern Illinois University.

Russell J. Cox, Executive Vice President and Chief Commercial Officer

Mr. Cox was appointed Executive Vice President and Chief Commercial Officer of Parent as of March 2012 and served as its Senior Vice President, Sales and Marketing from the Azur Merger until March 2012. Prior to the Azur Merger, he served in a variety of senior management roles since joining Jazz Pharmaceuticals, Inc. in 2010. From January 2009 to January 2010, he was Senior Vice President and Chief Commercial Officer of Ipsen Group, and from 2007 until December 2008, he was Vice President of Marketing at Tercica, Inc. (acquired by Ipsen Group), a biotechnology company. From 2003 to 2007, he was with Scios Inc. (acquired by Johnson and Johnson later in 2003), where he also held the role of Vice President, Marketing. Prior to 2003, Mr. Cox was with Genentech, Inc. for 12 years, where he was a Product Team Leader (PTL) responsible for the Growth Hormone franchise and led numerous product launches as a Group Product Manager. Mr. Cox received a B.S. in Biomedical Science from Texas A&M University.

Kathryn E. Falberg, Executive Vice President and Chief Financial Officer

Ms. Falberg was appointed Executive Vice President and Chief Financial Officer of Parent as of March 2012 and served as its Senior Vice President and Chief Financial Officer from the Azur Merger until March 2012. Prior to the Azur Merger, she served as Jazz Pharmaceuticals, Inc.'s Senior Vice President and Chief Financial Officer since joining Jazz Pharmaceuticals, Inc. in December 2009. From 2003 to 2008, Ms. Falberg was President of Canyon Capital & Consulting, a private investment and consulting firm, where she worked with a number of

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NAME AND POSITION	PRESENT PRINCIPAL OCCUPATION OR EMPLOYMENT AND EMPLOYMENT HISTORY
	smaller companies while also serving as a corporate director and audit committee chair for several companies, and from February to November 2009, she was Chief Financial Officer and Chief Operating Officer at ARCA biopharma, Inc., a biopharmaceutical company. From 1995 through 2001, Ms. Falberg was with Amgen, Inc., where she served as Senior Vice President Finance, Strategy and Chief Financial Officer, and before that as Vice President, Controller and Chief Accounting Officer, and Vice President, Treasurer. Ms. Falberg received an M.B.A. and B.A. in Economics from the University of California, Los Angeles and is a Certified Public Accountant (inactive). Ms. Falberg currently serves on the boards, and is chair of the audit committees, of the biopharmaceutical companies Halozyme Therapeutics, Inc. and Medivation, Inc.
Fintan Keegan, Executive Vice President, Technical Operations	Please see Mr. Keegan's biography set forth in Section 1 of this Schedule I.
Suzanne Sawochka Hooper, Executive Vice President and General Counsel	Ms. Hooper was appointed Executive Vice President and General Counsel of Parent as of March 2012. From 1999 through early 2012, she was a partner in the law firm Cooley LLP. Ms. Hooper served for several years as a member of Cooley's Management Committee and as Vice Chair of the firm's Business Department. While at Cooley, Ms. Hooper practiced corporate and securities law, primarily with companies and investors in the life sciences industry. Ms. Hooper received a J.D. from the University of California, Berkeley, Boalt Hall School of Law and a B.A. in Political Science from the University of California, Santa Barbara. Ms. Hooper is a member of the State Bar of California.
Jeffrey K. Tobias, M.D., Executive Vice President, Research and Development and Chief Medical Officer	Dr. Tobias was appointed Executive Vice President, Research and Development and Chief Medical Officer of Parent as of March 2012 and served as its Senior Vice President, Research and Development and Chief Medical Officer from the Azur Merger until March 2012. Prior to the Azur Merger, he served as Jazz Pharmaceuticals, Inc.'s Senior Vice President, Research and Development and Chief Medical Officer since joining Jazz Pharmaceuticals, Inc. in October 2011. From January 2010 to October 2011, Dr. Tobias served as Executive Vice President, Research and Development at NeurogesX, Inc.; previously, he served as NeurogesX's Chief Medical Officer since November 2005. Dr. Tobias was founder and managing director of the Aquila Consulting Group, LLC, a biopharmaceutical consulting firm, from September 1996 to November 2005. Prior to these activities, Dr. Tobias was a Director, New Product Discovery at ALZA Corporation, Director, Clinical Development at Chiron Corporation and Director, Clinical Research at Xoma Corporation. Dr. Tobias received board certification in both Internal Medicine and Pulmonary Medicine and completed training in Critical Care Medicine at the University of California, Los Angeles. He received an M.D. with honors and a B.A. from the University of Illinois.



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**NAME AND POSITION**

**PRESENT PRINCIPAL OCCUPATION OR  
EMPLOYMENT AND EMPLOYMENT HISTORY**

Karen J. Wilson, Senior Vice President, Finance  
and Principal Accounting Officer

Ms. Wilson has served as Senior Vice President, Finance and Principal Accounting Officer of Parent since February 2013 and served as its Vice President, Finance and Principal Accounting Officer from the Azur Merger until February 2013. Prior to the Azur Merger, she served as Jazz Pharmaceuticals, Inc.'s Vice President, Finance since February 2011 and was appointed Principal Accounting Officer in March 2011. From 2009 to January 2011, Ms. Wilson served as Vice President of Finance and Principal Accounting Officer at PDL BioPharma, Inc. From 2005 to 2009, she served as a principal at the consulting firm Wilson Crisler LLC. Previously, from 2001 to 2004, she was Chief Financial Officer of ViroLogic, Inc. Prior to joining ViroLogic, Ms. Wilson served as Chief Financial Officer and Vice President of Operations for Novare Surgical Systems, Inc. from 1999 to 2001. Prior to 1999, Ms. Wilson worked for Deloitte & Touche LLP for ten years, serving clients in both the medical and technology fields. Ms. Wilson is a Certified Public Accountant (inactive) in the State of California and received a B.S. in Business from the University of California, Berkeley.

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Manually signed facsimiles of the Share Form of Acceptance or ADS Letter of Transmittal, properly completed, will be accepted. The Share Form of Acceptance, ADS Letter of Transmittal, the ADRs and any other required documents should be sent by each holder or such holder's broker other securities intermediary to the Tender Agent at one of its addresses set forth below:

The Tender Agent for the Offer is:

**THE BANK OF NEW YORK MELLON**

***By registered, certified or express mail:***

The Bank of New York Mellon  
Voluntary Corporate Actions  
P. O. Box 3031  
Providence, Rhode Island 04940-3031  
United States of America

***By overnight courier:***

The Bank of New York Mellon  
Voluntary Corporate Actions – Suite V  
250 Royall Street  
Canton, Massachusetts 02021  
United States of America

Questions or requests for assistance may be directed to the Information Agent at the address and telephone numbers listed below. Additional copies of this Offer to Purchase, the Share Form of Acceptance, the ADS Letter of Transmittal and the Notice of Guaranteed Delivery may also be obtained from the Information Agent. Holders of ADSs may also contact brokers and other securities intermediaries for assistance concerning the Offer.

*The Information Agent for the Offer is:*

**MACKENZIE  
PARTNERS, INC.**

105 Madison Avenue  
New York, New York 10016  
(212) 929-5500 (Call Collect)

or

(800) 322-2885 (Toll Free)

Email: [tenderoffer@mackenziepartners.com](mailto:tenderoffer@mackenziepartners.com)

**THIS DOCUMENT IS IMPORTANT AND REQUIRES YOUR IMMEDIATE ATTENTION.**

**ADS Letter of Transmittal**  
**To Tender American Depositary Shares**  
**(each American Depositary Share representing one Ordinary Share)**  
**of**  
**GENTIUM S.p.A.**  
**CUSIP: 37250B14**  
**at**  
**\$57.00 Per American Depositary Share**  
**Pursuant to the Offer to Purchase dated December 23, 2013**  
**by**  
**JAZZ PHARMACEUTICALS ITALY S.r.l.**  
**a wholly-owned subsidiary of**  
**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

**THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.**

***The Tender Agent for the Offer is:***  
**The Bank of New York Mellon**

*By Registered, Certified or Express Mail:*  
The Bank of New York Mellon  
Voluntary Corporate Actions  
P.O. Box 3031  
Providence, Rhode Island 04940-3031  
United States of America

*By Overnight Courier:*  
The Bank of New York Mellon  
Voluntary Corporate Actions – Suite V  
250 Royall Street  
Canton, Massachusetts 02021  
United States of America

***The Information Agent for the Offer is:***

***MACKENZIE  
PARTNERS, INC***

105 Madison Avenue  
New York, New York 10016  
(212) 929-5500 (Call Collect)  
or  
(800) 322-2885 (Toll Free)  
Email: tenderoffer@mackenziepartners.com

VOLUNTARY CORPORATE ACTIONS COY: GENB

**DESCRIPTION OF ADSs TENDERED**

Name(s) and Address(es) of Registered Owner(s) (If blank, please fill in exactly as name(s) appear(s) on ADR(s))	ADSs Tendered (Attach additional list if necessary)		
	ADR Number(s)*	Total Number of ADSs Represented By ADR(s)*	Number of ADSs Tendered**
	<b>Total ADSs</b>		

- \* Need not be completed by holders tendering ADSs by book-entry transfer.
- \*\* Unless otherwise indicated, it will be assumed that all ADSs represented by ADRs delivered to the Tender Agent are being tendered hereby. See Instruction 4.
- \*\*\* If any of the ADRs that you own have been lost, destroyed, mutilated or stolen, see Instruction 10. If any ADRs have been lost, destroyed, mutilated or stolen, please fill out the remainder of this ADS Letter of Transmittal and indicate here the number of ADSs represented by the lost, destroyed, mutilated or stolen ADR(s) please call 1-888-BNY ADRS.

**DELIVERY OF THIS ADS LETTER OF TRANSMITTAL AND THE GENTIUM ADR CERTIFICATE(S) TO AN ADDRESS OTHER THAN AS SET FORTH ABOVE WILL NOT CONSTITUTE A VALID DELIVERY. YOU MUST SIGN THIS ADS LETTER OF TRANSMITTAL WHERE INDICATED BELOW AND, IF YOU ARE A U.S. HOLDER, COMPLETE THE SUBSTITUTE FORM W-9 ACCOMPANYING THIS ADS LETTER OF TRANSMITTAL OR IRS FORM W-9; IF YOU ARE A NON-U.S. HOLDER, YOU MUST COMPLETE A FORM W-8BEN OR OTHER FORM W-8, AS APPLICABLE.**

**PLEASE READ THE INSTRUCTIONS ACCOMPANYING THIS ADS LETTER OF TRANSMITTAL CAREFULLY BEFORE COMPLETING THIS ADS LETTER OF TRANSMITTAL.**

You have received this ADS Letter of Transmittal in connection with the offer by Jazz Pharmaceuticals Italy S.r.l., an Italian limited liability company (“**Purchaser**”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“**Parent**”), to purchase all outstanding shares of ordinary stock, no par value per share (the “**Ordinary Shares**”), and all outstanding American Depositary Shares, each representing one Ordinary Share (“**ADSs**”), of Gentium S.p.A., a *società per azioni* incorporated in Italy (“**Gentium**”), for \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the tendering holder in cash, without interest thereon and less any required withholding taxes, as described in the Offer to Purchase, dated December 23, 2013 (the “**Offer to Purchase**”). This document should be read in conjunction with the Offer to Purchase.

You should use this ADS Letter of Transmittal to deliver to The Bank of New York Mellon as tender agent for the Offer (as defined below) (the “**Tender Agent**”) ADSs represented by American Depositary Receipts (“**ADRs**”) for tender. If you hold your ADSs in book-entry form, you must request your broker or other securities intermediary to effect the transaction for you. Ordinary Shares cannot be tendered by means of this ADS Letter of Transmittal for ADSs. If you are a holder of Ordinary Shares you can obtain a Share Form of Acceptance and related documents for tendering those securities from the Information Agent. This ADS Letter of Transmittal for

ADSs can be used only to tender ADSs. See the Instructions on this ADS Letter of Transmittal for ADSs. If you are delivering your ADSs by book-entry transfer to an account maintained by the Tender Agent at The Depository Trust Company (“DTC”), you must instruct your broker or other securities intermediary in a timely manner to use an Agent’s Message (as defined in Instruction 2 below). In this document, holders who deliver ADRs representing their ADSs are referred to as “ADR Holders.” **Delivery of documents to an address other than one of those set forth for the Tender Agent above will not constitute delivery to the Tender Agent.**

If ADRs for your ADSs are not immediately available or you cannot deliver your ADRs and all other required documents to the Tender Agent on or prior to the Expiration Date (as defined below) of the Offer, or you cannot comply with the book-entry transfer procedures on a timely basis, you may nevertheless tender your ADSs according to the guaranteed delivery procedures set forth in the section “THE TENDER OFFER – Procedures for Tendering Ordinary Shares or ADSs” of the Offer to Purchase. See Instruction 2. The term “**Expiration Date**” means 12:00 Midnight, New York City time, on the evening of January 22, 2014, or if the Offer is extended, the latest time and date at which the Offer, as extended, will expire.

**THE OFFER HAS NOT BEEN SUBMITTED FOR CLEARANCE TO CONSOB (THE ITALIAN SECURITIES AND EXCHANGE COMMISSION). ACCORDINGLY, THE OFFER IS NOT BEING MADE AND WILL NOT BE MADE, DIRECTLY OR INDIRECTLY, IN ITALY, AND NO COPIES OF THIS DOCUMENT, THE OFFER TO PURCHASE OR ANY OTHER DOCUMENT RELATING TO THE OFFER HAVE BEEN OR WILL BE DISTRIBUTED IN ITALY, EXCEPT THAT, AS REQUIRED BY LAW, HOLDERS OF RECORD OR THOSE APPEARING ON A SECURITIES POSITION LISTING AS HOLDERS OF ADSs (INCLUDING, PURSUANT TO AN EXEMPTION AVAILABLE UNDER ITALIAN LAW, THE VERY LIMITED NUMBER OF SUCH HOLDERS LOCATED OR RESIDENT IN ITALY) WILL BE SENT COPIES OF THIS DOCUMENT AND OTHER DOCUMENTS RELATING TO THE OFFER. NEITHER THIS DOCUMENT NOR THE OFFER TO PURCHASE NOR ANY OTHER DOCUMENT RELATING TO THE OFFER MAY BE DISTRIBUTED TO ANY OTHER PERSON LOCATED OR RESIDENT IN ITALY FOR ANY REASON. NEITHER ANY SUCH DOCUMENT NOR ANY INFORMATION CONTAINED HEREIN OR THEREIN CONSTITUTES AN OFFER TO PURCHASE OR A SOLICITATION OF AN OFFER TO SELL OR AN ADVERTISEMENT OF AN OFFER TO PURCHASE ADSs IN ITALY WITHIN THE MEANING OF ARTICLE 1, PARAGRAPH 1, LETTER (V) OF ITALIAN LEGISLATIVE DECREE N. 58 OF FEBRUARY 24, 1998.**

**CHECK HERE IF TENDERED ADSs ARE BEING DELIVERED PURSUANT TO A NOTICE OF GUARANTEED DELIVERY PREVIOUSLY SENT TO THE TENDER AGENT AND COMPLETE THE FOLLOWING. PLEASE ENCLOSE A PHOTOCOPY OF SUCH NOTICE OF GUARANTEED DELIVERY.**

Name(s) of Registered Owner(s): \_\_\_\_\_

DTC Participant Number: \_\_\_\_\_

Date of Execution of Notice of Guaranteed Delivery: \_\_\_\_\_

Name of Institution which Guaranteed Delivery: \_\_\_\_\_

Transaction Code Number: \_\_\_\_\_

**NOTE: SIGNATURES MUST BE PROVIDED BELOW**  
**PLEASE READ THE ACCOMPANYING INSTRUCTIONS CAREFULLY**

Ladies and Gentlemen:

The undersigned hereby tenders to Purchaser the above-described ADSs pursuant to the Offer to Purchase, at a price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, on the terms and subject to the conditions set forth in the Offer to Purchase, receipt of which is hereby acknowledged, and this ADS Letter of Transmittal (which, together with the Offer to Purchase and the Share Form of Acceptance, as amended or supplemented from time to time, collectively constitute the “Offer”).

The undersigned understands that the acceptance for payment by Purchaser of ADSs tendered pursuant to one of the procedures described in the section “THE TENDER OFFER – Procedures for Tendering Ordinary Shares or ADSs” of the Offer to Purchase will constitute a binding agreement between the undersigned and Purchaser, upon the terms and subject to the conditions of the Offer.

On the terms and subject to the conditions of the Offer (including, if the Offer is extended or amended, the terms and conditions of such extension or amendment) and to the extent permitted under applicable law, subject to, and effective upon, acceptance for payment and payment for the ADSs validly tendered herewith in accordance with the terms of the Offer, the undersigned hereby sells, assigns and transfers to, or upon the order of, Purchaser, all right, title and interest in and to all of the and ADSs being tendered hereby and any and all cash dividends, distributions, rights, other Ordinary Shares, ADSs or other securities issued or issuable in respect of such ADSs on or after December 23, 2013 (collectively, “Distributions”). In addition, the undersigned irrevocably appoints the Tender Agent as the true and lawful agent and attorney-in-fact of the undersigned with respect to such ADSs and any Distributions with full power of substitution (such power of attorney being deemed to be an irrevocable power coupled with an interest) to the fullest extent of the undersigned’s rights with respect to such ADSs and any Distributions to (a) deliver ADRs representing ADSs and any Distributions, and to transfer ownership of such ADSs and any Distributions on the account books maintained by DTC, together, in either such case, with all accompanying evidence of transfer and authenticity, to or upon the order of Purchaser, (b) present such ADSs and any Distributions for transfer on the books of Gentium and (c) receive all benefits and otherwise exercise all rights of beneficial ownership of such ADSs and any Distributions, all in accordance with the terms and subject to the conditions of the Offer and to the extent permitted under applicable law.

To the extent permitted under applicable law, the undersigned hereby irrevocably appoints each of the designees of Purchaser the attorneys-in-fact and proxies of the undersigned, each with full power of substitution, to the full extent of the undersigned’s rights with respect to the ADSs tendered hereby that have been accepted for payment and with respect to any Distributions. The designees of Purchaser will, with respect to the ADSs and any associated Distributions for which the appointment is effective, be empowered to exercise all voting and any other rights of the undersigned, as they, in their sole discretion, may deem proper. This proxy and power of attorney shall be irrevocable and coupled with an interest in the tendered ADSs. Such appointment is effective when, and only to the extent that, Purchaser accepts the ADSs tendered with this ADS Letter of Transmittal for payment pursuant to the Offer. Upon the effectiveness of such appointment, without further action, all prior powers of attorney, proxies and consents given by the undersigned with respect to such ADSs and any associated Distributions will be revoked and no subsequent powers of attorney, proxies, consents or revocations may be given (and, if given, will not be deemed effective). Purchaser reserves the right to require that, in order for ADSs to be deemed validly tendered, immediately upon Purchaser’s acceptance for payment of such ADSs, Purchaser must be able to exercise full voting, consent and other rights, to the extent permitted under applicable law, with respect to such ADSs and any associated Distributions, including voting at any meeting of shareholders concerning any matter.

The undersigned hereby represents and warrants that the undersigned has full power and authority to tender, sell, assign and transfer the ADSs and any Distributions tendered hereby and, when the same are accepted for

payment by Purchaser, Purchaser will acquire good, marketable and unencumbered title thereto, free and clear of all liens, restrictions, charges and encumbrances and the same will not be subject to any adverse claim. The undersigned hereby represents and warrants that the undersigned is the registered owner of the ADSs, or that any ADR(s) have been endorsed to the undersigned in blank or the undersigned is a participant in DTC whose name appears on a security position listing as the owner of the ADSs. The undersigned will, upon request, execute and deliver any additional documents deemed by the Tender Agent or Purchaser to be necessary or desirable to complete the sale, assignment and transfer of the ADSs and any Distributions tendered hereby. In addition, the undersigned shall promptly remit and transfer to the Tender Agent for the account of Purchaser any and all Distributions in respect of the ADSs tendered hereby, accompanied by appropriate documentation of transfer and, pending such remittance or appropriate assurance thereof, Purchaser shall be entitled to all rights and privileges as owner of any such Distributions and may withhold the entire purchase price or deduct from the purchase price the amount or value thereof, as determined by Purchaser in its sole discretion.

It is understood that the undersigned will not receive payment for the ADSs unless and until the ADSs are accepted for payment and until any ADR(s) owned by the undersigned are received by the Tender Agent at the address set forth above, together with such additional documents as the Tender Agent may require, or, in the case of ADSs held in book-entry form, ownership of ADSs is validly transferred on the account books maintained by DTC, and until the same are processed for payment by the Tender Agent. It is understood that the method of delivery of the ADR(s) and all other required documents is at the option and risk of the undersigned and will be deemed made only when actually received by the Tender Agent. In all cases, sufficient time should be allowed to ensure timely delivery. Registered mail with return receipt requests or overnight courier properly insured is recommended for ADRs evidencing ADSs sent by mail.

All authority conferred or agreed to be conferred pursuant to this ADS Letter of Transmittal shall not be affected by, and shall survive, the death or incapacity of the undersigned and any obligation of the undersigned hereunder shall be binding upon the heirs, executors, administrators, trustees in bankruptcy, personal representatives, successors and assigns of the undersigned. Except as stated in the Offer to Purchase, this tender is irrevocable.

Unless otherwise indicated herein under "Special Payment Instructions," please issue the check for the purchase price and/or return any ADRs representing ADSs not tendered or accepted for payment in the name(s) of the registered owner(s) appearing under "Description of ADSs Tendered." Similarly, unless otherwise indicated under "Special Delivery Instructions," please mail the check for the purchase price and/or return any ADRs representing ADSs not tendered or accepted for payment (and accompanying documents, as appropriate) to the address(es) of the registered owner(s) appearing under "Description of ADSs Tendered." In the event that both the Special Delivery Instructions and the Special Payment Instructions are completed, please issue the check for the purchase price and/or issue any ADRs representing ADSs not tendered or accepted for payment (and any accompanying documents, as appropriate) in the name of, and deliver such check and/or return such ADRs (and any accompanying documents, as appropriate) to, the person or persons so indicated. Unless otherwise indicated herein in the box titled "Special Payment Instructions," please credit any ADSs tendered hereby or by an Agent's Message and delivered by book-entry transfer, but which are not purchased, by crediting the account at DTC designated above. The undersigned recognizes that Purchaser has no obligation pursuant to the Special Payment Instructions to transfer any ADSs from the name of the registered owner thereof if Purchaser does not accept for payment any of the ADSs so tendered.

**SPECIAL PAYMENT INSTRUCTIONS**  
**(See Instructions 1, 5, 6 and 7)**

To be completed ONLY if ADR(s) representing ADSs not tendered or not accepted for payment and/or the check for the purchase price of ADSs accepted for payment (less any required withholding taxes) are to be issued in the name of someone other than the undersigned or if ADSs tendered by book-entry transfer which are not accepted for payment are to be returned by credit to an account maintained at DTC other than that designated above.

Issue:  Check and/or  
 ADRs to:

Name: \_\_\_\_\_  
**(Please Print)**

Address: \_\_\_\_\_

\_\_\_\_\_  
**(Include Zip Code)**

\_\_\_\_\_  
**(Tax Identification or Social Security Number)**

Credit ADSs tendered by book-entry transfer that are not accepted for payment to the DTC account set forth below.

\_\_\_\_\_  
**(DTC Account Number)**

**SPECIAL DELIVERY INSTRUCTIONS**

To be completed ONLY if ADR(s) representing ADSs not tendered or not accepted for payment and/or the check for the purchase price of ADSs accepted for payment (less any required withholding taxes) are to be sent to someone other than the undersigned or to the undersigned at an address other than that shown in the box titled "Description of ADSs Tendered" above.

Deliver:  Check and/or  
 ADRs to:

Name: \_\_\_\_\_  
**(Please Print)**

Address: \_\_\_\_\_

\_\_\_\_\_  
**(Include Zip Code)**

\_\_\_\_\_  
**(Tax Identification or Social Security Number)**



**IMPORTANT – SIGN HERE**  
**(U.S. Holders Please Also Complete Substitute Form W-9 Included Herein)**  
**(Non-U.S. Holders Please Obtain and Complete Form W-8BEN or Other Form W-8)**

(all holders must sign)

\_\_\_\_\_  
\_\_\_\_\_  
(Signature(s) of Holder(s))

Dated: \_\_\_\_\_, 20\_\_

(Must be signed by registered owner(s) exactly as name(s) appear(s) on ADR(s) or on a security position listing or by person(s) authorized to become registered owner(s) by certificates and documents transmitted herewith. If signature is by trustees, executors, administrators, guardians, attorneys-in-fact, officers of corporations or others acting in a fiduciary or representative capacity, please set forth full title and see Instruction 5.)

Name(s): \_\_\_\_\_  
\_\_\_\_\_

\_\_\_\_\_  
\_\_\_\_\_  
(Please Print)

Capacity (full title): \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

Address: \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

\_\_\_\_\_  
(Include Zip Code)

Area Code and Telephone Number: \_\_\_\_\_

Tax Identification or Social Security No.: \_\_\_\_\_

**GUARANTEE OF SIGNATURE(S)**  
**(For use by Eligible Institutions only; see Instructions 1 and 5)**

Name of Firm: \_\_\_\_\_

Address: \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

\_\_\_\_\_  
(Include Zip Code)

Authorized Signature: \_\_\_\_\_

Name: \_\_\_\_\_

\_\_\_\_\_  
(Please Type or Print)

Area Code and Telephone Number: \_\_\_\_\_

Dated: \_\_\_\_\_, 20\_\_

**Place medallion guarantee in space below:**

## INSTRUCTIONS

### Forming Part of the Terms and Conditions of the Offer

1. **Guarantee of Signatures.** Except as otherwise provided below, all signatures on this ADS Letter of Transmittal must be guaranteed by a financial institution (including most commercial banks, savings and loan associations and brokerage houses) that is a member in good standing of a recognized Medallion Program approved by the Securities Transfer Association, Inc., including the Security Transfer Agents Medallion Program, the New York Stock Exchange Medallion Signature Program and the Stock Exchange Medallion Program (each, an “**Eligible Institution**”). Medallion guarantees from Eligible Institutions are generally not available outside the United States. If you need and are unable to obtain a medallion guarantee from an Eligible Institution, please contact the Information Agent for instructions. See Instruction 5. Signatures on this ADS Letter of Transmittal need not be guaranteed (a) if this ADS Letter of Transmittal is signed by the registered owner(s) (which term, for purposes of this document, includes any participant in DTC whose name appears on a security position listing as the owner of the ADSs) of ADSs tendered herewith and such registered owner has not completed either the box titled “Special Payment Instructions” or the box titled “Special Delivery Instructions” on this ADS Letter of Transmittal or (b) if such ADSs are tendered for the account of an Eligible Institution.

2. **Delivery of ADS Letter of Transmittal and ADRs or Book-Entry Confirmations.** This ADS Letter of Transmittal is to be used either if ADRs are to be forwarded herewith or, unless an Agent’s Message is utilized, if tenders are to be made pursuant to the procedures for tender by book-entry transfer set forth in the section “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs” of the Offer to Purchase. An originally signed facsimile of this document may be used in lieu of the original. ADRs representing all physically tendered ADSs, or confirmation of any book-entry transfer into the Tender Agent’s account at DTC of ADSs tendered by book-entry transfer (“**Book Entry Confirmation**”), as well as this ADS Letter of Transmittal properly completed and duly executed with any required signature guarantees, or an Agent’s Message in the case of a book-entry transfer, and any other documents required by this ADS Letter of Transmittal, must be received by the Tender Agent at one of its addresses set forth herein on or prior to the Expiration Date (as described in the section “THE TENDER OFFER – Terms of the Offer” of the Offer to Purchase). Please do not send your ADRs directly to Purchaser, Parent or Gentium.

Holders whose ADRs are not immediately available or who cannot deliver all other required documents to the Tender Agent on or prior to the Expiration Date or who cannot comply with the procedures for book-entry transfer on a timely basis, may nevertheless tender their ADSs by properly completing and duly executing a Notice of Guaranteed Delivery pursuant to the guaranteed delivery procedure set forth in the section “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs” of the Offer to Purchase. Pursuant to such procedure: (a) such tender must be made by or through an Eligible Institution, (b) a properly completed and duly executed Notice of Guaranteed Delivery substantially in the form provided by Purchaser must be received by the Tender Agent prior to the Expiration Date and (c) ADRs representing all tendered ADSs, in proper form for transfer (or a Book-Entry Confirmation with respect to ADSs), as well as a ADS Letter of Transmittal (or an originally signed facsimile thereof), properly completed and duly executed with any required signature guarantees (or, in the case of a book-entry transfer, an Agent’s Message in lieu of a ADS Letter of Transmittal), and all other documents required by this ADS Letter of Transmittal, must be received by the Tender Agent within three Nasdaq Global Select Market trading days after the date of execution of such Notice of Guaranteed Delivery. See Instruction 5.

A properly completed and duly executed ADS Letter of Transmittal (or an originally signed facsimile thereof) must accompany each such delivery of ADRs to the Tender Agent.

The term “**Agent’s Message**” means a message, transmitted by DTC to, and received by, the Tender Agent and forming part of a Book-Entry Confirmation, which states that DTC has received an express acknowledgment from the participant in DTC tendering the ADSs which are the subject of such Book-Entry Confirmation, that such participant has received and agrees to be bound by the terms of the ADS Letter of Transmittal and that Purchaser may enforce such agreement against the participant.

**THE METHOD OF DELIVERY OF THE ADSs, THIS ADS LETTER OF TRANSMITTAL AND ALL OTHER REQUIRED DOCUMENTS, INCLUDING DELIVERY THROUGH DTC, IS AT THE ELECTION AND RISK OF THE TENDERING HOLDER. DELIVERY OF ALL SUCH DOCUMENTS WILL BE DEEMED MADE AND RISK OF LOSS OF THE ADRs SHALL PASS ONLY WHEN ACTUALLY RECEIVED BY THE TENDER AGENT (INCLUDING, IN THE CASE OF A BOOK-ENTRY TRANSFER, BY BOOK-ENTRY CONFIRMATION). IF SUCH DELIVERY IS BY MAIL, IT IS RECOMMENDED THAT ALL SUCH DOCUMENTS BE SENT BY PROPERLY INSURED REGISTERED MAIL WITH RETURN RECEIPT REQUESTED. IN ALL CASES, SUFFICIENT TIME SHOULD BE ALLOWED TO ENSURE TIMELY DELIVERY.**

No alternative, conditional or contingent tenders will be accepted and no fractional ADSs will be purchased. All tendering holders, by execution of this ADS Letter of Transmittal (or an originally signed facsimile thereof), waive any right to receive any notice of the acceptance of their ADSs for payment.

All questions as to validity, form, eligibility (including time of receipt) and acceptance for payment of any tender of ADSs hereunder will be determined by Purchaser, in its sole discretion (which may delegate power in whole or in part to the Tender Agent) and such determination shall be final and binding. Purchaser reserves the right to waive any irregularities or defects in the surrender of any ADSs. A surrender will not be deemed to have been made until all irregularities have been cured or waived. None of Purchaser, Parent or any of their respective affiliates or assigns, the Tender Agent, the Information Agent (as defined in the Offer to Purchase), or any other person will be under any duty to give notification of any defects or irregularities in tenders or incur any liability for failure to give any such notification. Purchaser's interpretation of the terms and conditions of the Offer (including this ADS Letter of Transmittal and the instructions hereto and any other documents related to the Offer) will be final and binding.

**3. Inadequate Space.** If the space provided herein is inadequate, the certificate numbers and/or the number of ADSs should be listed on a separate schedule attached hereto and separately signed on each page thereof in the same manner as this ADS Letter of Transmittal is signed.

**4. Partial Tenders (Applicable to ADR Holders Only).** If fewer than all ADSs evidenced by any ADR delivered to the Tender Agent are to be tendered, fill in the number of ADSs which are to be tendered in the box titled "Number of ADSs Tendered." In such cases, new ADRs for the remainder of the ADSs that were evidenced by the old ADRs but not tendered will be sent to the registered owner, unless otherwise provided in the appropriate box on this ADS Letter of Transmittal, as soon as practicable after the Expiration Date. All ADSs represented by ADRs delivered to the Tender Agent will be deemed to have been tendered unless otherwise indicated.

**5. Signatures on ADS Letter of Transmittal; Stock Powers and Endorsements.** If this ADS Letter of Transmittal is signed by the registered owner(s) of the ADSs tendered hereby, the signature(s) must correspond with the name(s) as written on the face of the ADRs without alteration, enlargement or any other change whatsoever.

If any of the ADSs tendered hereby are owned of record by two or more joint owners, all such owners must sign this ADS Letter of Transmittal.

If any of the tendered ADSs are registered in the names of different holder(s), it will be necessary to complete, sign and submit as many separate ADS Letters of Transmittal (or originally signed facsimiles thereof) as there are different registrations of such ADSs.

If this ADS Letter of Transmittal or any certificates or stock powers are signed by trustees, executors, administrators, guardians, attorneys-in-fact, officers of corporations or others acting in a fiduciary or representative capacity, such persons should so indicate when signing, and proper evidence satisfactory to Purchaser of their authority so to act must be submitted.

If this ADS Letter of Transmittal is signed by the registered owner(s) of the ADSs listed and transmitted hereby, no endorsements of ADRs or separate stock powers are required unless payment is to be made to, or ADRs representing ADSs not tendered or accepted for payment are to be issued in the name of, a person other than the registered owner(s). Signatures on such ADRs or stock powers must be guaranteed by an Eligible Institution. See Instruction 1.

If this ADS Letter of Transmittal is signed by a person other than the registered owner(s) of the ADS(s) listed, the ADR(s) must be endorsed or accompanied by the appropriate stock powers, in either case, signed exactly as the name or names of the registered owner(s) or holder(s) appear(s) on the ADR(s). Signatures on such ADRs or stock powers must be guaranteed by an Eligible Institution.

**6. Stock Transfer Taxes.** Except as otherwise provided in this Instruction 6, Purchaser will pay any stock transfer taxes with respect to the transfer and sale of ADSs to it or to its order pursuant to the Offer. If, however, payment of the purchase price is to be made to, or (in the circumstances permitted hereby) if ADRs representing ADSs not tendered or accepted for payment are to be registered in the name of, any person resident in Italy other than the registered owner(s), or if tendered ADRs are registered in the name of any person resident in Italy other than the person signing this ADS Letter of Transmittal, any such registered owner(s) or person shall independently establish whether it owes any stock transfer taxes in connection with such transfer and, in case any such taxes shall be due, pay them directly to the competent authorities.

**Except as provided in this Instruction 6, it will not be necessary for transfer tax stamps to be affixed to the ADRs listed in this ADS Letter of Transmittal.**

**7. Special Payment and Delivery Instructions.** If a check is to be issued in the name of, and/or ADRs representing ADSs not tendered or accepted for payment are to be issued or returned to, a person other than the signer(s) of this ADS Letter of Transmittal or if a check and/or such certificates are to be mailed to a person other than the signer(s) of this ADS Letter of Transmittal or to an address other than that shown in the box titled "Description of ADSs Tendered" above, the appropriate boxes on this ADS Letter of Transmittal should be completed. Holders delivering ADSs tendered hereby or by Agent's Message by book-entry transfer may request that such ADSs not purchased be credited to an account maintained at DTC as such shareholder may designate in the box titled "Special Payment Instructions" herein. If no such instructions are given, all such ADSs not purchased will be returned by crediting the same account at DTC as the account from which such ADSs were delivered.

**8. Requests for Assistance or Additional Copies.** Questions or requests for assistance may be directed to the Information Agent at its address and telephone number set forth below or to your broker or other securities intermediary. Additional copies of the Offer to Purchase, this ADS Letter of Transmittal, the Notice of Guaranteed Delivery and other tender offer materials may be obtained from the Information Agent as set forth below, subject to applicable law, and will be furnished at Purchaser's expense.

**9. Backup Withholding.** In order to avoid U.S. federal backup withholding with respect to cash received in exchange for ADSs pursuant to the Offer, a holder who is a U.S. person submitting ADSs must (i) provide the Tender Agent with a properly completed Substitute Form W-9, included in this ADS Letter of Transmittal, or IRS Form W-9 and sign such form under penalties of perjury, (ii) provide the Tender Agent with a properly completed IRS Form W-8BEN or other Form W-8, and sign such form under penalties of perjury, or (iii) otherwise establish an exemption. IRS Form W-9, and W-8BEN and other Forms W-8 (if you are not a U.S. person) are available from the Tender Agent or from the Internal Revenue Service web site, at <http://www.irs.ustreas.gov>.

**10. Lost, Destroyed, Mutilated or Stolen ADRs.** If any ADR has been lost, destroyed, mutilated or stolen, the holder should complete, sign and return to the Tender Agent this ADS Letter of Transmittal indicating the number of ADRs that have been lost, destroyed, mutilated or stolen, or call the Tender Agent at 1-888-BNY-ADRS for further instructions.

11. **Waiver of Conditions.** Subject to the terms and conditions of the Offer to Purchase and the applicable rules and regulations of the Securities and Exchange Commission, Purchaser reserves the right to waive in whole or in part at any time and from time to time in its sole discretion any of the conditions of the Offer prior to expiration of the Offer.

**IMPORTANT: THIS ADS LETTER OF TRANSMITTAL (OR AN ORIGINALLY SIGNED FACSIMILE THEREOF) OR AN AGENT'S MESSAGE, TOGETHER WITH ADRs OR BOOK-ENTRY CONFIRMATION OR A PROPERLY COMPLETED AND DULY EXECUTED NOTICE OF GUARANTEED DELIVERY AND ALL OTHER REQUIRED DOCUMENTS MUST BE RECEIVED BY THE TENDER AGENT ON OR PRIOR TO THE EXPIRATION DATE.**

## IMPORTANT TAX INFORMATION

Under the U.S. federal income tax law, a holder who is a U.S. person whose tendered ADSs are accepted for purchase is required by law to (i) provide the Tender Agent with such holder's correct taxpayer identification number ("TIN") on Substitute Form W-9 below, certify that such TIN is correct (or that such security holder is awaiting a TIN), and certify whether the holder is subject to backup withholding or (ii) otherwise establish a basis for exemption from backup withholding. If such holder of ADSs is an individual, the TIN is his or her social security number. If a holder of ADSs fails to provide a correct TIN to the Tender Agent, such person may be subject to a U.S.\$50.00 penalty imposed by the Internal Revenue Service. In addition, payments that are made to such holder of ADSs with respect to ADSs purchased pursuant to the Offer may be subject to backup withholding of 28%.

Certain holders of ADSs (including, among others, all corporations and certain foreign individuals) are not subject to these backup withholding and reporting requirements. In order for a foreign person to qualify for an exemption from backup withholding, that holder must generally submit a Form W-8BEN (or other applicable Form W-8), signed under penalties of perjury, attesting to that individual's exempt status. A Form W-8BEN can be obtained from the Tender Agent or the IRS website, at <http://www.irs.ustreas.gov>.

If backup withholding applies, the Tender Agent is required to withhold 28% of any payments made to the Ordinary Share or ADS holder or payee. Backup withholding is not an additional tax. Rather, the U.S. federal income tax liability of persons subject to backup withholding will be reduced by the amount of tax withheld. If withholding results in an overpayment of taxes, a refund may be obtained from the Internal Revenue Service by timely filing a claim for refund.

If backup withholding applies and "Applied For" is written in Part I of the Substitute Form W-9 and the holder of ADSs has completed the Certificate of Awaiting Taxpayer Identification Number, the Tender Agent will retain 28% of any payment of the purchase price for tendered ADSs during the 60-day period following the date of the Substitute Form W-9. If a holder's TIN is provided to the Tender Agent within 60 days of the date of the Substitute Form W-9, payment of such retained amounts will be made to such holder of ADSs. If a holder's TIN is not provided to the Tender Agent within such 60-day period, the Tender Agent will remit such retained amounts to the Internal Revenue Service as backup withholding and shall withhold 28% of any payment of the purchase price for the tendered ADSs made to such holder thereafter unless such holder of ADSs furnishes a TIN to the Tender Agent prior to such payment.

### **Purpose of Substitute Form W-9**

To prevent backup withholding on payments made to a holder whose tendered ADSs are accepted for purchase (for holders other than foreign persons who provide an appropriate Form W-8BEN), the holder should complete and sign the Substitute Form W-9 included in this ADS Letter of Transmittal and provide the holder's correct TIN and certify, under penalties of perjury, that the TIN provided on such Form is correct (or that such holder is awaiting a TIN) and that (a) such holder of ADSs is exempt from backup withholding, (b) such holder of ADSs has not been notified by the Internal Revenue Service that such holder is subject to backup withholding as a result of failure to report all interest or dividends or (c) the Internal Revenue Service has notified the holder that the holder is no longer subject to backup withholding. The holder must sign and date the Substitute Form W-9 where indicated, certifying that the information on such Form is correct.

### **What Number to Give the Tender Agent on the Substitute Form W-9**

The holder of ADSs is required to give the Tender Agent the social security number or employer identification number of the record owner of the ADSs. If the ADSs are in more than one name or are not in the name of their actual holder, consult the enclosed Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9 for additional guidance on which number to report.

**TO BE COMPLETED BY ALL TENDERING U.S. HOLDERS**  
**(See Instruction 9)**  
**PAYER: The Bank of New York Mellon**

<p><b>SUBSTITUTE</b></p> <p><b>Form W-9</b></p> <p><b>Request for Taxpayer Identification Number (TIN) and Certification</b></p>	<p>Name: _____</p> <p>Address: _____</p> <p>_____</p> <p>Check appropriate box:</p> <table style="width:100%; border:none;"> <tr> <td style="width:50%;">Individual/Sole Proprietor <input type="checkbox"/></td> <td>Limited Liability Company. Enter the tax classification:</td> </tr> <tr> <td>S Corporation <input type="checkbox"/></td> <td>C corporation <input type="checkbox"/></td> </tr> <tr> <td>C Corporation <input type="checkbox"/></td> <td>S corporation <input type="checkbox"/></td> </tr> <tr> <td>Trust/Estate <input type="checkbox"/></td> <td>Partnership <input type="checkbox"/></td> </tr> <tr> <td>Partnership <input type="checkbox"/></td> <td></td> </tr> <tr> <td>Other (specify) <input type="checkbox"/></td> <td></td> </tr> </table>	Individual/Sole Proprietor <input type="checkbox"/>	Limited Liability Company. Enter the tax classification:	S Corporation <input type="checkbox"/>	C corporation <input type="checkbox"/>	C Corporation <input type="checkbox"/>	S corporation <input type="checkbox"/>	Trust/Estate <input type="checkbox"/>	Partnership <input type="checkbox"/>	Partnership <input type="checkbox"/>		Other (specify) <input type="checkbox"/>	
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Trust/Estate <input type="checkbox"/>	Partnership <input type="checkbox"/>												
Partnership <input type="checkbox"/>													
Other (specify) <input type="checkbox"/>													
<p><b>Part I.</b> Please provide your taxpayer identification number in the space at right. If awaiting TIN, write "Applied For" in space at right and complete the Certificate of Awaiting Taxpayer Identification Number below.</p>													
<table style="width:100%; border:none;"> <tr> <td style="width:75%;"></td> <td style="width:25%;"><b>SSN:</b></td> </tr> <tr> <td></td> <td>Or</td> </tr> <tr> <td></td> <td><b>EIN:</b> _____</td> </tr> </table>			<b>SSN:</b>		Or		<b>EIN:</b> _____						
	<b>SSN:</b>												
	Or												
	<b>EIN:</b> _____												
<p><b>Part II.</b> For Payees exempt from backup withholding, see the enclosed "Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9" and complete as instructed therein.</p>													
<p><b>Part III. CERTIFICATION</b></p> <p>Under penalties of perjury, I certify that:</p> <p>(1) The number shown on this form is my correct Taxpayer Identification Number (or, as indicated, I am waiting for a number to be issued to me);</p> <p>(2) I am not subject to backup withholding because: (a) I am exempt from backup withholding, or (b) I have not been notified by the IRS that I am subject to backup withholding as a result of a failure to report all interests or dividends, or (c) the IRS has notified me that I am no longer subject to backup withholding; and</p> <p>(3) I am a U.S. citizen or other U.S. person (including a U.S. resident alien).</p> <p>Certification Instructions – You must cross out item (2) above if you have been notified by the IRS that you are subject to backup withholding because of underreporting of interest or dividends on your tax return. However, if after being notified by the IRS that you were subject to backup withholding you received another notification from the IRS that you are no longer subject to backup withholding, do not cross out item (2).</p>													
<p>Signature of U.S. person: _____ Date: _____</p>													

**YOU MUST COMPLETE THE FOLLOWING CERTIFICATE IF YOU WROTE "APPLIED FOR"  
IN PART I OF THIS SUBSTITUTE FORM W-9**

**CERTIFICATE OF AWAITING TAXPAYER IDENTIFICATION NUMBER**

I certify under penalties of perjury that a taxpayer identification number has not been issued to me, and either (a) I have mailed or delivered an application to receive a taxpayer identification number to the appropriate Internal Revenue Service Center or Social Security Administration Office or (b) I intend to mail or deliver an application in the near future. I understand that, notwithstanding the information I provided in Part III of the Substitute Form W-9 (and the fact that I have completed this Certificate of Awaiting Taxpayer Identification Number), all reportable payments made to me hereafter will be subject to a 28 percent backup withholding tax until I provide a properly certified taxpayer identification number.

\_\_\_\_\_  
Signature

\_\_\_\_\_  
Date

**NOTE: FAILURE TO COMPLETE AND RETURN THIS FORM MAY RESULT IN BACKUP WITHHOLDING OF 28% OF ANY PAYMENTS MADE TO YOU PURSUANT TO THIS OFFER. PLEASE REVIEW THE ENCLOSED GUIDELINES FOR CERTIFICATION OF TAXPAYER IDENTIFICATION NUMBER ON SUBSTITUTE FORM W-9 FOR ADDITIONAL DETAILS.**



***The Tender Agent for the Offer is:  
The Bank of New York Mellon***

*By Registered, Certified or Express Mail:*  
The Bank of New York Mellon  
Voluntary Corporate Actions  
P.O. Box 3031  
Providence, Rhode Island 04940-3031  
United States of America

*By Overnight Courier:*  
The Bank of New York Mellon  
Voluntary Corporate Actions – Suite V  
250 Royall Street  
Canton, Massachusetts 02021  
United States of America

***The Information Agent for the Offer is:***

***MACKENZIE  
PARTNERS, INC***

105 Madison Avenue  
New York, New York 10016  
(212) 929-5500 (Call Collect)  
or  
(800) 322-2885 (Toll Free)  
Email: [tenderoffer@mackenziepartners.com](mailto:tenderoffer@mackenziepartners.com)

December 23, 2013

VOLUNTARY CORPORATE ACTIONS COY: GENB

## Notice of Guaranteed Delivery

**for Offer to Purchase for Cash  
All Outstanding Ordinary Shares and American Depositary Shares  
(each American Depositary Share representing one Ordinary Share)**

of

**GENTIUM S.p.A.**

at

**\$57.00 per Ordinary Share and  
per American Depositary Share**

by

**JAZZ PHARMACEUTICALS ITALY S.r.l.**

a wholly-owned subsidiary of

**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

**THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF  
JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.**

As set forth under “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs – Guaranteed Delivery” in the Offer to Purchase, dated December 23, 2013 (as may from time to time be amended, supplemented or finalized, the “Offer to Purchase”), this Notice of Guaranteed Delivery, or a form substantially equivalent hereto, must be used to accept the offer of Jazz Pharmaceuticals Italy S.r.l., an Italian limited liability company (“Purchaser”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“Parent”), to purchase all outstanding shares of ordinary stock, no par value per share (“Ordinary Shares”) of Gentium S.p.A., a *società per azioni* incorporated in Italy (“Gentium”) and all outstanding American Depositary Shares (“ADSs”), each representing one Ordinary Share and evidenced by an American Depositary Receipt (an “ADR”) at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer to Purchase, and in the related ADS Letter of Transmittal (which, together with the Offer to Purchase and the Form of Share Acceptance, each as may be amended or supplemented from time to time, collectively constitute the “Offer”) if:

- certificates evidencing ADRs representing ADSs are not immediately available;
- the procedures for book-entry transfer of ADSs cannot be completed prior to the Expiration Date of the Offer; or
- time will not permit all required documents to reach The Bank of New York Mellon, as the tender agent for the Offer (the “Tender Agent”), prior to the Expiration Date.

The term “Expiration Date” means 12:00 Midnight, New York City Time, on the evening of January 22, 2014, unless extended, in which event the term “Expiration Date” shall mean the latest time and date at which the Offer, as so extended, will expire. This Notice of Guaranteed Delivery may be delivered by facsimile transmission, mail or overnight courier to the Tender Agent **and must include a Guarantee by an Eligible Institution** (as defined in the ADS Letter of Transmittal). See the guaranteed delivery procedures described in the Offer to Purchase under “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs – Guaranteed Delivery.”

VOLUNTARY CORPORATE ACTIONS COY: GENB

*The Tender Agent for the Offer is:*  
**THE BANK OF NEW YORK MELLON**

***By registered, certified or express mail:***

The Bank of New York Mellon  
Voluntary Corporate Actions  
P. O. Box 3031  
Providence, Rhode Island 04940-3031  
United States of America  
Facsimile: (617) 360 - 6810  
For Confirmation Only/Telephone Number: (781) 575-2332

***By overnight courier:***

The Bank of New York Mellon  
Voluntary Corporate Actions – Suite V  
250 Royall Street  
Canton, Massachusetts 02021  
United States of America

**DELIVERY OF THIS NOTICE OF GUARANTEED DELIVERY TO AN ADDRESS OTHER THAN ONE SET FORTH ABOVE OR TRANSMISSION OF INSTRUCTIONS VIA FACSIMILE TO A NUMBER OTHER THAN THE FACSIMILE NUMBER SET FORTH ABOVE DOES NOT CONSTITUTE A VALID DELIVERY.**

THIS NOTICE OF GUARANTEED DELIVERY TO THE TENDER AGENT IS NOT TO BE USED TO GUARANTEE SIGNATURES. IF A SIGNATURE ON AN ADS LETTER OF TRANSMITTAL IS REQUIRED TO BE GUARANTEED BY AN ELIGIBLE INSTITUTION UNDER THE INSTRUCTIONS THERETO, SUCH SIGNATURE GUARANTEE MUST APPEAR IN THE APPLICABLE SPACE PROVIDED IN THE SIGNATURE BOX ON THE ADS LETTER OF TRANSMITTAL.

THE GUARANTEE INCLUDED HEREIN MUST BE COMPLETED.

VOLUNTARY CORPORATE ACTIONS COY: GENB

**Ladies and Gentlemen:**

The undersigned represents that the undersigned owns and hereby tenders to Jazz Pharmaceuticals Italy S.r.l., an Italian limited liability company (“Purchaser”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“Parent”), upon the terms and subject to the conditions set forth in the Offer to Purchase, dated December 23, 2013 (as may from time to time be amended, supplemented or finalized, the “Offer to Purchase”) and in the related ADS Letter of Transmittal (together with the Form of Share Acceptance and any amendments or supplements thereto, collectively constitute the “Offer”), receipt of which is hereby acknowledged, the number of ADSs set forth below, all pursuant to the guaranteed delivery procedures set forth in the Offer to Purchase.

Name(s) of Record Holder(s):

Number of ADSs Tendered:

ADR Number (if available):

Address(es):

Name of Tendering Institution:

Area Code and Telephone No.(s):

Signature(s):

Check if ADSs will be book-entry transfer

Name of Transferring Institution:

DTC Account No.:

Transaction Code No.:

Dated:

VOLUNTARY CORPORATE ACTIONS COY: GENB

**GUARANTEE**  
**(Not to be used for signature guarantee)**

The undersigned, a financial institution that is a participant in the Security Transfer Agent Medallion Program, or any other "eligible guarantor institution," as such term is defined in Rule 17Ad-15 under the Securities Exchange Act of 1934, as amended, hereby guarantees to deliver to the Tender Agent: either (i) the ADRs representing the ADSs tendered hereby, in proper form for transfer, or (ii) to deliver a timely confirmation of a book-entry transfer of such ADSs into the Tender Agent's account at The Depository Trust Company ("DTC"), in any such case together with a properly completed and duly executed ADS Letter of Transmittal, with any required signature guarantees or, in the case of a book-entry transfer of such ADSs in the Tender Agent's account at DTC, an Agent's Message (as defined in the Offer to Purchase) and any other documents required by the ADS Letter of Transmittal, all within three trading days after the date hereof.

The Eligible Institution that completes this form must communicate the guarantee to the Tender Agent and must deliver the properly completed and duly executed ADS Letter of Transmittal or an Agent's Message and certificates representing the ADRs representing ADSs to the Tender Agent within the time period shown herein. Failure to do so could result in a financial loss to such Eligible Institution.

Name of Firm: \_\_\_\_\_

Address: \_\_\_\_\_  
(Zip Code)

Area Code and Tel. No. \_\_\_\_\_

Name: \_\_\_\_\_  
(Please type or print)

\_\_\_\_\_  
(Authorized Signature)

Title: \_\_\_\_\_

Dated: \_\_\_\_\_

**NOTE: DO NOT SEND CERTIFICATES FOR ADRs REPRESENTING ADSs WITH THIS NOTICE. CERTIFICATES FOR ADRs REPRESENTING ADSs SHOULD BE SENT WITH YOUR PROPERLY COMPLETED AND DULY EXECUTED ADS LETTER OF TRANSMITTAL.**

**Offer To Purchase For Cash**  
**All Outstanding Ordinary Shares and American Depositary Shares**  
 (each American Depositary Share representing one Ordinary Share)  
 of  
**GENTIUM S.P.A.**  
 at  
**\$57.00 per ORDINARY SHARE and per AMERICAN DEPOSITARY SHARE**  
 by  
**JAZZ PHARMACEUTICALS ITALY S.r.l.**  
 a wholly-owned subsidiary of  
**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

<b>THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF          JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.</b>
--

December 23, 2013

**To Brokers, Dealers, Commercial Banks, Trust Companies and Other Nominees:**

Enclosed for your consideration are the Offer to Purchase, dated December 23, 2013 (as may from time to time be amended, supplemented or finalized, the "Offer to Purchase"), and the related Share Form of Acceptance and ADS Letter of Transmittal (which, together with any amendments or supplements thereto, collectively constitute the "Offer") in connection with the offer by Jazz Pharmaceuticals Italy S.r.l., an Italian limited liability company ("Purchaser") and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland ("Parent"), to purchase all outstanding shares of ordinary stock, no par value per share ("Ordinary Shares") of Gentium S.p.A., a *società per azioni* incorporated in Italy ("Gentium") and all outstanding American Depositary Shares ("ADSs"), each representing one Ordinary Share and evidenced by an American Depositary Receipt (an "ADR") at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer.

The Offer is being made pursuant to a Tender Offer Agreement, dated as of December 19, 2013 (as it may be amended from time to time, the "Tender Offer Agreement"), by and among Parent, Purchaser and Gentium.

Please furnish copies of the enclosed materials to those of your clients, **other than those resident in Italy**, for whose accounts you hold ADSs registered in your name or in the name of your nominee.

Enclosed herewith are copies of the following documents:

1. The Offer to Purchase, dated December 23, 2013.
2. The Share form of Acceptance to be used to tender Ordinary Shares for your use and for the information of your clients, together with the included Internal Revenue Service Form W-9.
3. The ADS Letter of Transmittal to be used to tender ADSs for your use and for the information of your clients, together with the included Internal Revenue Service Form W-9.
3. The Notice of Guaranteed Delivery to be used to accept the Offer if the procedures for tendering ADSs set forth in the Offer to Purchase and the ADS Letter of Transmittal cannot be completed prior to January 22, 2014, at 12:00 Midnight, New York City time, unless the Offer is extended (the "Expiration Date").
4. A printed form of letter which may be sent to your clients for whose accounts you hold Ordinary Shares and/or ADSs registered in your name or in the name of your nominee, with space provided for obtaining such clients' instructions with regard to the Offer.

5. A copy of Gentium's Solicitation/Recommendation Statement on Schedule 14D-9, which has been filed by Gentium with the Securities and Exchange Commission (the "SEC").
6. A return envelope addressed to The Bank of New York Mellon, Voluntary Corporate Actions – Suite V, P. O. Box 3031, Providence, Rhode Island 04940-3031 (the "Tender Agent").

Consummation of the Offer is subject to various conditions set forth in the Tender Offer Agreement, including, but not limited to the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Ordinary Shares and ADSs that, together with any Ordinary Shares and ADSs owned by Parent or Purchaser or any of their subsidiaries, represents at least 66 2/3% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the time that Purchaser first accepts any Ordinary Shares and ADSs for payment pursuant to the Offer (the "Acceptance Time"), plus (B) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time in all cases without duplication for Ordinary Shares underlying the ADSs provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time, (without duplication for Ordinary Shares underlying ADSs) (the "Permitted Minimum Condition")

**WE URGE YOU TO CONTACT YOUR CLIENTS (OTHER THAN THOSE THAT ARE RESIDENT IN ITALY) AS PROMPTLY AS POSSIBLE. PLEASE NOTE THAT THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.**

**The Company's Board of Directors (the "Company Board") has determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company's shareholders and other stakeholders of the Company. The Company Board has approved and deemed advisable the Tender Offer Agreement and the transactions contemplated thereby and has recommended that the holders of Ordinary Shares and ADSs accept the Offer and tender their Ordinary Shares and ADSs to Purchaser pursuant to the Offer.**

For purposes of the Offer, Purchaser will be deemed to have accepted for payment and thereby purchased Ordinary Shares and ADSs validly tendered and not properly withdrawn if and when Purchaser gives notice to The Bank of New York Mellon (the "Tender Agent") of its acceptance for payment of such Ordinary Shares and ADSs pursuant to the Offer. Payment for Ordinary Shares and ADSs accepted for payment pursuant to the Offer will be made by deposit of the purchase price therefor with the Tender Agent, which will act as agent for the tendering holders for purposes of receiving payments from Purchaser and transmitting such payments to the tendering holders. If Purchaser does not accept Ordinary Shares pursuant to the Offer, Ordinary Shares tendered by book-entry transfer into the Monte Titoli account of the Tender Agent's Custodian (as defined in the Offer to Purchase) pursuant to the procedures set forth under "The Offer – Procedure for Tendering Ordinary Shares and ADSs" section of the Offer to Purchase will be credited to the account maintained with Monte Titoli from which those shares were originally transferred, promptly following expiration or termination of the Offer. If Purchaser does not accept ADSs for exchange pursuant to the Offer or if more ADRs are submitted for ADSs than are tendered in the Offer, Purchaser will return ADRs for these unexchanged ADSs without expense to the tendering stockholder. If Purchaser does not accept ADSs pursuant to the Offer, ADSs tendered by book – entry transfer into the Tender Agent's account at The Depository Trust Company ("DTC") pursuant to the procedures set forth under "The Offer – Procedure for Tendering Ordinary Shares and ADSs" section of the Offer to Purchase will be credited to the account maintained with DTC from which those shares were originally transferred, promptly following expiration or termination of the Offer. UNDER NO CIRCUMSTANCES WILL INTEREST BE PAID ON THE OFFER PRICE FOR TENDERED ORDINARY SHARES OR ADSs, REGARDLESS OF ANY EXTENSION OF THE OFFER OR ANY DELAY IN MAKING PAYMENT.

The Offer is not being made to holders of Ordinary Shares or ADSs in any jurisdiction in which the making or acceptance of the Offer would not be in compliance with the laws of such jurisdiction or any administrative or judicial action pursuant thereto. However, Purchaser may, in its discretion, take such action as it may deem necessary to make the Offer in any jurisdiction and extend the Offer to holders of Ordinary Shares and ADSs in such jurisdiction. **FOR THE AVOIDANCE OF DOUBT, THE OFFER HAS NOT BEEN SUBMITTED FOR CLEARANCE TO CONSOB (THE ITALIAN SECURITIES AND EXCHANGE COMMISSION). ACCORDINGLY, THE OFFER IS NOT BEING MADE AND WILL NOT BE MADE, DIRECTLY OR INDIRECTLY, IN ITALY AND NO COPIES OF THE OFFER TO PURCHASE OR ANY OTHER DOCUMENT RELATING TO THE OFFER HAVE BEEN OR WILL BE DISTRIBUTED IN ITALY, EXCEPT THAT, AS REQUIRED BY LAW, HOLDERS OF RECORD OR THOSE APPEARING ON A SECURITIES POSITION LISTING AS HOLDERS OF ADSS OR ORDINARY SHARES (INCLUDING, PURSUANT TO AN EXEMPTION AVAILABLE UNDER ITALIAN LAW, THE VERY LIMITED NUMBER OF SUCH HOLDERS LOCATED OR RESIDENT IN ITALY) WILL BE SENT COPIES OF CERTAIN DOCUMENTS RELATING TO THE OFFER. NEITHER THIS DOCUMENT NOR THE OFFER TO PURCHASE NOR ANY OTHER DOCUMENT RELATING TO THE OFFER MAY BE DISTRIBUTED TO ANY OTHER PERSON LOCATED OR RESIDENT IN ITALY FOR ANY REASON. NEITHER ANY SUCH DOCUMENT NOR ANY INFORMATION CONTAINED HEREIN OR THERIN CONSTITUTES AN OFFER TO PURCHASE OR A SOLICITATION OF AN OFFER TO SELL OR AN ADVERTISEMENT OF AN OFFER TO PURCHASE ORDINARY SHARES OR ADSs IN ITALY WITHIN THE MEANING OF ARTICLE 1, PARAGRAPH 1, LETTER (V) OF ITALIAN LEGISLATIVE DECREE N. 58 OF FEBRUARY 24, 1998.**

Neither Parent nor Purchaser nor any officer, director, stockholder, agent or other representative of Parent or Purchaser will pay any fees or commissions to any broker or dealer or other person (other than to the Tender Agent or the Information Agent) for soliciting tenders of Ordinary Shares and ADSs in the Offer.

Purchaser will, however, upon request, reimburse brokers, dealers, commercial banks and trust companies for customary mailing and handling costs incurred by them in forwarding the enclosed materials to their customers.

In order to take advantage of the Offer, (i) ADSs or certificates evidencing ADRs representing ADSs into the Tender Agent's account at DTC pursuant to the procedures set forth in "THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs" of the Offer to Purchase, as well as an ADS Letter of Transmittal, properly completed and duly executed with any required signature guarantees (or, in the case of a book-entry transfer, an Agent's Message (as defined in the Offer to Purchase) in lieu of the ADS Letter of Transmittal), and all other documents required by the ADS Letter of Transmittal must be received by the Tender Agent, all in accordance with the instructions set forth in the ADS Letter of Transmittal and the Offer to Purchase or (ii) a timely book-entry transfer of Ordinary Shares to the Monte Titoli account of the Tender Agent's Custodian (as defined in the Offer to Purchase) pursuant to the procedures set forth in "THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs" of the Offer to Purchase, as well as a Share Form of Acceptance, properly completed and duly executed, and all other documents required by the Share Form of Acceptance must be received by the Tender Agent, all in accordance with the instructions set forth in the Share Form of Acceptance and the Offer to Purchase.

Any holder of ADSs who desires to tender such ADSs whose certificate(s) evidencing such ADRs representing such ADSs are not immediately available, or cannot comply with the procedures for book-entry transfer described in the Offer to Purchase on a timely basis may tender such ADSs by following the procedures for guaranteed delivery set forth in "THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs – Guaranteed Delivery" of the Offer to Purchase.

Any inquiries you may have with respect to the Offer should be addressed to MacKenzie Partners, Inc., the information agent for the Offer (the "Information Agent"), by mail at 105 Madison Avenue, New York, New York 10016, or by phone at (212) 929-5500 (collect) or (800) 322-2885 (toll-free).



Requests for copies of the enclosed materials may also be directed to the Information Agent at the above telephone number.

Very truly yours,

MacKenzie Partners, Inc.

**Nothing contained herein or in the enclosed documents shall constitute you the agent of Parent, Purchaser, Gentium, the Information Agent, the Tender Agent or any affiliate of any of the foregoing, or authorize you or any other person to use any document or make any statement on behalf of any of them in connection with the offer other than the documents enclosed herewith and the statements contained therein.**

**Offer To Purchase For Cash**  
**All Outstanding Ordinary Shares and American Depositary Shares**  
 (each American Depositary Share representing one Ordinary Share)  
 of  
**GENTIUM S.p.A.**  
 at  
**\$57.00 per ORDINARY SHARE and per AMERICAN DEPOSITARY SHARE**  
 by  
**JAZZ PHARMACEUTICALS ITALY S.r.l.,**  
 a wholly-owned subsidiary of  
**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

<p><b>THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.</b></p>
--

December 23, 2013

**To Our Clients:**

Enclosed for your consideration are the Offer to Purchase, dated December 23, 2013 (as may from time to time be amended, supplemented or finalized, the "Offer to Purchase"), and the related Share Form of Acceptance and ADS Letter of Transmittal (which, together with any amendments or supplements thereto, collectively constitute the "Offer") in connection with the offer by Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* ("Purchaser") and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland ("Parent"), to purchase all outstanding shares of ordinary stock, no par value per share ("Ordinary Shares") of Gentium S.p.A., a *società per azioni* incorporated in Italy ("Gentium") and all outstanding American Depositary Shares ("ADSs"), each representing one Ordinary Share and evidenced by an American Depositary Receipt (an "ADR") at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer. Also enclosed is Gentium's Solicitation/Recommendation Statement on Schedule 14D-9.

The Offer is being made pursuant to a Tender Offer Agreement, dated as of December 19, 2013 (as it may be amended from time to time, the "Tender Offer Agreement"), by and among Parent, Purchaser and Gentium.

**We are (or our nominee is) the holder of record of the Ordinary Shares and/or ADSs held for your account. A tender of such Ordinary Shares and/or ADSs can be made only by us as the holder of record and pursuant to your instructions. The enclosed Share Form of Acceptance and Letter of Transmittal are furnished to you for your information only and cannot be used by you to tender such Ordinary Shares and/or ADSs held by us for your account.**

Accordingly, we request instructions as to whether you wish us to tender on your behalf any or all of the Ordinary Shares and/or ADSs held by us for your account, upon the terms and subject to the conditions set forth in the Offer. Your attention is directed to the following:

1. The consideration per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs) is \$57.00, net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer.
2. The Offer is being made for all outstanding Ordinary Shares and ADSs.
3. **The Company's Board of Directors (the "Company Board") has determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company's shareholders and other stakeholders of the Company. The Company Board has approved and deemed advisable the Tender Offer Agreement and the transactions**

contemplated thereby and has recommended that the holders of Ordinary Shares and ADSs accept the Offer and tender their Ordinary Shares and ADSs to Purchaser pursuant to the Offer.

**4. THE OFFER AND WITHDRAWAL RIGHTS EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED (THE “EXPIRATION DATE”).**

Consummation of the Offer is subject to various conditions set forth in the Tender Offer Agreement, including, but not limited to the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Ordinary Shares and ADSs that, together with any Ordinary Shares and ADSs owned by Parent or Purchaser or any of their subsidiaries, represents at least 66 2/3% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the time that Purchaser first accepts any Ordinary Shares and ADSs for payment pursuant to the Offer (the “Acceptance Time”), plus (B) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time in all cases without duplication for Ordinary Shares underlying the ADSs provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying ADSs) (the “Permitted Minimum Condition”).

If you wish to have us tender any of the Ordinary Shares and/or ADSs held by us for your account, please instruct us by completing, executing and returning to us in the enclosed envelope the instruction form contained in this letter. If you authorize a tender of your Ordinary Shares and/or ADSs, all such Ordinary Shares and/or ADSs will be tendered unless otherwise specified in such instruction form. **Your instructions should be forwarded to us in ample time to permit us to submit a tender on your behalf prior to the Expiration Date.**

Payment for Ordinary Shares accepted for payment in the Offer will in all cases be made only after timely receipt by the Tender Agent of: (i) a timely book-entry transfer of such Ordinary Shares to the Monte Titoli account of the Tender Agent’s Custodian (as defined in the Offer to Purchase) pursuant to the procedures set forth in “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs” of the Offer to Purchase; (ii) a properly completed and duly executed Share Form of Acceptance (or an originally signed facsimile thereof); and (iii) all other documents required by the Share Form of Acceptance. Payment for ADSs accepted for payment in the Offer will in all cases be made only after timely receipt by the Tender Agent of: (i) certificates evidencing ADRs representing such ADSs or a timely Book-Entry Confirmation (as defined in the Offer to Purchase) of such ADSs into the Tender Agent’s account at the Depository Trust Company (“DTC”) pursuant to the procedures set forth in “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs” of the Offer to Purchase; (ii) a properly completed and duly executed ADS Letter of Transmittal (or an originally signed facsimile thereof), with any required signature guarantees (or, in the case of a book-entry transfer effected pursuant to the procedures set forth in “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs” of the Offer to Purchase, an Agent’s Message (as defined in the Offer to Purchase) in lieu of an ADS Letter of Transmittal); and (iii) all other documents required by the ADS Letter of Transmittal. **UNDER NO CIRCUMSTANCES WILL INTEREST BE PAID ON THE OFFER PRICE FOR ORDINARY SHARES OR ADSs, REGARDLESS OF ANY EXTENSION OF THE OFFER OR ANY DELAY IN MAKING PAYMENT.**

The Offer is not being made to holders of Ordinary Shares or ADSs in any jurisdiction in which the making or acceptance of the Offer would not be in compliance with the laws of such jurisdiction or any administrative or judicial action pursuant thereto. However, Purchaser may, in its discretion, take such action as it may deem necessary to make the Offer in any jurisdiction and extend the Offer to holders of Ordinary Shares and ADSs in such jurisdiction. **FOR THE AVOIDANCE OF DOUBT, THE OFFER HAS NOT BEEN SUBMITTED**

FOR CLEARANCE TO CONSOB (THE ITALIAN SECURITIES AND EXCHANGE COMMISSION). ACCORDINGLY, THE OFFER IS NOT BEING MADE AND WILL NOT BE MADE, DIRECTLY OR INDIRECTLY, IN ITALY AND NO COPIES OF THE OFFER TO PURCHASE OR ANY OTHER DOCUMENT RELATING TO THE OFFER HAVE BEEN OR WILL BE DISTRIBUTED IN ITALY, EXCEPT THAT, AS REQUIRED BY LAW, HOLDERS OF RECORD OR THOSE APPEARING ON A SECURITIES POSITION LISTING AS HOLDERS OF ADSS OR ORDINARY SHARES (INCLUDING, PURSUANT TO AN EXEMPTION AVAILABLE UNDER ITALIAN LAW, THE VERY LIMITED NUMBER OF SUCH HOLDERS LOCATED OR RESIDENT IN ITALY) WILL BE SENT COPIES OF CERTAIN DOCUMENTS RELATING TO THE OFFER. NEITHER THIS DOCUMENT NOR THE OFFER TO PURCHASE NOR ANY OTHER DOCUMENT RELATING TO THE OFFER MAY BE DISTRIBUTED TO ANY OTHER PERSON LOCATED OR RESIDENT IN ITALY FOR ANY REASON. NEITHER ANY SUCH DOCUMENT NOR ANY INFORMATION CONTAINED HEREIN OR THEREIN CONSTITUTES AN OFFER TO PURCHASE OR A SOLICITATION OF AN OFFER TO SELL OR AN ADVERTISEMENT OF AN OFFER TO PURCHASE ORDINARY SHARES OR ADSs IN ITALY WITHIN THE MEANING OF ARTICLE I, PARAGRAPH 1, LETTER (V) OF ITALIAN LEGISLATIVE DECREE N. 58 OF FEBRUARY 24, 1998.

**INSTRUCTIONS WITH RESPECT TO THE OFFER TO PURCHASE FOR CASH  
ALL OUTSTANDING ORDINARY SHARES AND AMERICAN DEPOSITARY SHARES**

(each American Depositary Share representing one Ordinary Share)

of  
**GENTIUM S.p.A**  
by

**JAZZ PHARMACEUTICALS ITALY S.r.l.**

a wholly-owned subsidiary of

**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

The undersigned acknowledge(s) receipt of your letter, the enclosed Offer to Purchase, dated December 23, 2013, and the related Share Form of Certificate and ADS Letter of Transmittal (which, as they may be amended and supplemented from time to time, together constitute the "Offer") in connection with the offer Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* ("Purchaser") and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland ("Parent"), to purchase all outstanding shares of ordinary stock, no par value per share ("Ordinary Shares") of Gentium, a *società per azioni* incorporated in Italy ("Gentium") and all outstanding American Depositary Shares ("ADSs"), each representing one Ordinary Share and evidenced by an American Depositary Receipt (an "ADR") at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer.

This will instruct you to tender to Purchaser the number of Ordinary Shares and/or ADSs indicated below (or, if no number is indicated below, all Ordinary Shares and/or ADSs) which are held by you for the account of the undersigned, upon the terms and subject to the conditions set forth in the Offer.

The undersigned understand(s) and acknowledge(s) that all questions as to validity, form and eligibility of the surrender of any Ordinary Shares and/or ADSs submitted on my/our behalf to the Tender Agent (as defined in the Offer to Purchase) will be determined by Purchaser (which may delegate power in whole or in part to the Tender Agent) and such determination shall be final and binding.

**NUMBER OF ADSs TO BE TENDERED:**

**SIGN HERE**

\_\_\_\_\_ ADSs

\_\_\_\_\_  
\_\_\_\_\_  
(Signature(s))

**NUMBER OF ORDINARY SHARES  
TO BE TENDERED**

\_\_\_\_\_ Ordinary Shares

\_\_\_\_\_  
\_\_\_\_\_  
Please Type or Print Names(s)

\_\_\_\_\_  
Please Type or Print Names(s)

\_\_\_\_\_  
Tax Identification Number or Social Security Number

**Dated:** \_\_\_\_\_

**Please return this form to the brokerage firm or other nominee maintaining your account.**

**GUIDELINES FOR CERTIFICATION OF TAXPAYER IDENTIFICATION  
NUMBER ON SUBSTITUTE FORM W-9**

**Guidelines for Determining the Proper Identification Number for the Payee (You) to Give the Payor**—Social security numbers have nine digits separated by two hyphens (i.e., 000-00-0000). Employer identification numbers have nine digits separated by only one hyphen (i.e., 00-0000000). The table below will help determine the number to provide to the payer.

<b>For this type of account:</b>		<b>Give the name and social security number of:</b>	<b>For this type of account:</b>		<b>Give the name and taxpayer identification number of:</b>
1.	Individual	The individual	6.	Disregarded entity not owned by an individual	The owner
2.	Two or more individuals (joint account)	The actual owner of the account or, if combined funds, the first individual on the account <sup>1</sup>	7.	A valid trust, estate, or pension trust	The legal entity <sup>4</sup>
3.	Custodian account of a minor (Uniform Gift to Minors Act)	The minor <sup>2</sup>	8.	Corporate	The corporation
4(a)	The usual revocable savings trust (grantor is also trustee)	The grantor-trustee <sup>1</sup>	9.	Association, club, religious, charitable, educational, or other tax-exempt organization	The organization
4(b)	So-called trust account that is not a legal or valid trust under state law	The actual owner <sup>1</sup>	10.	Partnership	The partnership
5.	Sole proprietorship or disregarded entity not owned by an individual	The owner <sup>3</sup>	11.	A broker or registered nominee	The broker or nominee
			12.	Account with the Department of Agriculture in the name of a public entity (such as a state or local government, school district, or prison) that receives agricultural program payments	The public entity

- (1) List first and circle the name of the person whose number you furnish. If only one person on a joint account has a social security number, that person's number must be furnished.
- (2) Circle the minor's name and furnish the minor's social security number.
- (3) You must show your individual name, but you may also enter your business or "doing business as" name. You may use either your social security number or your taxpayer identification number (if you have one).
- (4) List first and circle the name of the legal trust, estate, or pension trust. (Do not furnish the taxpayer identification number of the personal representative or trustee unless the legal entity itself is not designated in the account title.)

**NOTE: If more than one name listed and no name is circled, the number will be considered to be that of the first name listed.**

# GUIDELINES FOR CERTIFICATION OF TAXPAYER IDENTIFICATION NUMBER ON SUBSTITUTE FORM W-9, Cont.

## Obtaining a Number

If you don't have a taxpayer identification number or you do not know your number, obtain Form SS-5, *Application for a Social Security Number Card*, or Form SS-4, *Application for Employer Identification Number*, at a local office of the Social Security Administration or the Internal Revenue Service or online at [www.ssa.gov](http://www.ssa.gov) or [www.irs.gov](http://www.irs.gov) and apply for a number. Section references in these guidelines refer to sections under the Internal Revenue Code of 1986, as amended.

## Payees Exempt From Backup Withholding

Even if the payee does not provide a taxpayer identification number in the manner required, you are **not required** to backup withhold on any payments you make if the payee is:

- An organization exempt from tax under Section 501(a), any individual retirement account (IRA), or a custodial account under Section 403(b)(7) if the account satisfies the requirements of Section 401(f)(2).
- The United States or any of its agencies or instrumentalities.
- A state, the District of Columbia, a possession of the United States, or any of their political subdivisions or instrumentalities.
- A foreign government or any of its political subdivisions, agencies, or instrumentalities.
- An international organization or any of its agencies or instrumentalities.

Other payees that **may be exempt** from backup withholding include:

- A corporation.
- A foreign central bank of issue.
- A dealer in securities or commodities required to register in the United States, the District of Columbia, or a possession of the United States.
- A futures commission merchant registered with the Commodity Futures Trading Commission.
- A real estate investment trust (REIT).
- An entity registered at all times during the tax year under the Investment Company Act of 1940.
- A common trust fund operated by a bank under Section 584(a).
- A financial institution.
- A middleman known in the investment community as a nominee or custodian.
- A trust exempt from tax under Section 664 or described in Section 4947.

## Payments Exempt From Backup Withholding

**Dividends and patronage dividends** that generally are exempt from backup withholding include:

- Payments to nonresident aliens subject to withholding under Section 1441.
- Payments to partnerships not engaged in a trade or business in the United States and that have at least one nonresident alien partner.
- Payments of patronage dividends not paid in money.
- Payments made by certain foreign organizations.
- Section 404(k) distributions made by an employee stock ownership plan (ESOP).

**Interest payments** that generally are exempt from backup withholding include:

- Payments of interest on obligations issued by individuals. However, if you pay \$600 or more of interest in the course of your trade or business to a payee, you must report the payment. Backup withholding applies to the reportable payment if the payee has not provided a taxpayer identification number or has provided an incorrect taxpayer identification number.
- Payments of tax-exempt interest (including exempt-interest dividends under Section 852).
- Payments described in Section 6049(b)(5) to nonresident aliens.
- Payments on tax-free covenant bonds under Section 1451.
- Payments made by certain foreign organizations.
- Mortgage or student loan interest paid to you.

**Other types of payments** that generally are exempt from backup withholding include:

- Wages.
- Distributions from a pension, annuity, profit-sharing or stock bonus plan, any IRA, an owner-employee plan or other deferred compensation plan.
- Distributions from a medical or health savings account and long-term care benefits.
- Certain surrenders of life insurance contracts.
- Gambling winnings if withholding is required under Section 3402(q). However, if withholding is not required under Section 3402(q), backup withholding applies if the payee fails to furnish a taxpayer identification number.
- Real estate transactions reportable under Section 6045(e).
- Cancelled debts reportable under Section 6050P.
- Fish purchases for cash reportable under Section 6050R.

Exempt payees described above should file Form W-9 to avoid possible erroneous backup withholding. **FILE THIS FORM WITH THE PAYER, FURNISH YOUR TAXPAYER IDENTIFICATION NUMBER, WRITE "EXEMPT" IN PART II OF THE FORM, SIGN AND DATE THE FORM AND RETURN IT TO THE PAYER. IF THE PAYMENTS ARE INTEREST, DIVIDENDS OR PATRONAGE DIVIDENDS, ALSO SIGN AND DATE THE FORM.**

Certain payments other than interest, dividends and patronage dividends not subject to information reporting are also not subject to backup withholding. For details, see the regulations under Sections 6041, 6041A, 6042, 6044, 6045, 6049, 6050A and 6050N of the Internal Revenue Code of 1986, as amended.

**Privacy Act Notice**—Section 6109 of the Internal Revenue Code of 1986, as amended, requires you to give your correct taxpayer identification number to persons who must file information returns with the Internal Revenue Service to report, among other things, interest, dividends, and certain other income paid to you. The Internal Revenue Service uses the numbers for identification purposes and to help verify the accuracy of your tax return. The Internal Revenue Service may also provide this information to the Department of Justice for civil and criminal litigation, and to cities, states and the District of Columbia to carry

## GUIDELINES FOR CERTIFICATION OF TAXPAYER IDENTIFICATION NUMBER ON SUBSTITUTE FORM W-9, Cont.

out their tax laws. You must provide your taxpayer identification number whether or not you are required to file a tax return. Payers must generally withhold 28% of taxable interest, dividend and certain other payments to a payee who does not give a taxpayer identification number to a payer. Certain penalties may also apply.

### Penalties—

**(1) Penalty for Failure to Furnish Taxpayer Identification Number**—If you fail to furnish your correct taxpayer identification number to a requester, you are subject to a penalty of \$50 for each such failure unless your failure is due to reasonable cause and not to willful neglect.

**(2) Civil Penalty for False Information With Respect to Withholding**—If you make a false statement with no reasonable basis that results in no imposition of backup withholding, you are subject to a penalty of \$500.

**(3) Civil and Criminal Penalties for False Information**—Willfully falsifying certifications or affirmations may subject you to criminal penalties including fines and/or imprisonment.

**(4) Misuse of Taxpayer Identification Numbers**—If the requester discloses or uses taxpayer identification numbers in violation of federal law, the requester may be subject to civil and criminal penalties.

**FOR ADDITIONAL INFORMATION CONTACT YOUR TAX  
CONSULTANT OR THE INTERNAL REVENUE SERVICE.**



**THIS DOCUMENT IS IMPORTANT AND REQUIRES YOUR IMMEDIATE  
ATTENTION.**

**Share Form of Acceptance  
To Tender Ordinary Shares  
of**

**GENTIUM S.p.A.**

ISIN: IT 0003747927

at

**\$57.00 Per Ordinary Share**

**Pursuant to the Offer to Purchase dated December 23, 2013**

by

**JAZZ PHARMACEUTICALS ITALY S.r.l.**

a wholly-owned subsidiary of

**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

**THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF  
JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED.**

***The Tender Agent for the Offer is:***  
**The Bank of New York Mellon**

*By Registered, Certified or Express Mail:*

The Bank of New York Mellon

Voluntary Corporate Actions

P.O. Box 3031

Providence, Rhode Island 04940-3031

United States of America

*By Overnight Courier:*

The Bank of New York Mellon

Voluntary Corporate Actions – Suite V

250 Royall Street

Canton, Massachusetts 02021

United States of America

VOLUNTARY CORPORATE ACTIONS COY: GENB

Facsimile: (617) 360-6810

For Confirmation Only/Telephone Number: (781) 575-2332

Remainder: The Signed Share Form of Acceptance, together with the completed tax form must be delivered to the Tender Agent prior to expiration of the Offer.

**THE INSTRUCTIONS IN THIS SHARE FORM OF ACCEPTANCE SHOULD BE READ PURSUANT TO THE TERMS AND CONDITIONS IN THE OFFER TO PURCHASE. TENDERS OF GENTIUM ADSs CANNOT BE MADE BY MEANS OF THIS SHARE FORM OF ACCEPTANCE.**

**PLEASE FILL IN THE INFORMATION BELOW WITH RESPECT TO THE ORDINARY SHARES BEING TENDERED BY BOOK-ENTRY TRANSFER VIA MONTE TITOLI TO THE TENDER AGENT'S ACCOUNT AT SOCIETE GENERALE:**

**Name of Tendering Institution: [Holder]:** \_\_\_\_\_

**Name of Monte Titoli Participant:** \_\_\_\_\_

**Contact Details of Monte Titoli Participant:** \_\_\_\_\_

**Transaction Number: [if applicable]** \_\_\_\_\_

**DESCRIPTION OF ORDINARY SHARES TENDERED**

Name(s), Address(es) and contact details of Registered Owner(s)*	Ordinary Shares Tendered

**DELIVERY OF THIS SHARE FORM OF ACCEPTANCE AND THE SIGNED TAX FORM TO AN ADDRESS OTHER THAN AS SET FORTH ABOVE WILL NOT CONSTITUTE A VALID DELIVERY. YOU MUST SIGN THIS SHARE FORM OF ACCEPTANCE WHERE INDICATED BELOW AND COMPLETE THE APPROPRIATE W-9 OR W-8 ACCOMPANYING THIS SHARE FORM OF ACCEPTANCE.**

**PLEASE READ THE INSTRUCTIONS ACCOMPANYING THIS SHARE FORM OF ACCEPTANCE CAREFULLY BEFORE COMPLETING THIS SHARE FORM OF ACCEPTANCE.**

You have received this Share Form of Acceptance in connection with the offer by Jazz Pharmaceuticals Italy S.r.l., an Italian limited liability company (“**Purchaser**”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“**Parent**”), to purchase all outstanding shares of ordinary stock, no par value per share (the “**Ordinary Shares**”), and all outstanding American Depositary Shares, each representing one Ordinary Share (“**ADSs**”), of Gentium S.p.A., a *società per azioni* incorporated in Italy (“**Gentium**”), for \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the tendering holder in cash, without interest thereon and less any required withholding taxes, as described in the Offer to Purchase, dated December 23, 2013 (the “**Offer to Purchase**”). This document should be read in conjunction with the Offer to Purchase.

You should use this Share Form of Acceptance to deliver to The Bank of New York Mellon as tender agent for the Offer (as defined in the Offer to Purchase) (the “**Tender Agent**”) Ordinary Shares held in book-entry form for tender. If you hold your Ordinary Shares in book-entry form, you must request your broker or other financial intermediary to effect the transaction for you. ADSs cannot be tendered by means of this Share Form of Acceptance for Ordinary Shares. If you are a holder of ADSs you can obtain an ADS Letter of Transmittal and related documents for tendering those securities from the Information Agent. This Share Form of Acceptance for Ordinary Shares can be used only to tender Ordinary Shares. If you are delivering your Ordinary Shares by book-entry transfer to an account maintained by the Tender Agent at *Société Générale*, you must instruct your securities intermediary in a timely manner to withdraw the Ordinary Shares from the account in Monte Titoli and arrange for the deposit of the Ordinary Shares in the Tender Agent’s account at *Société Générale*. This signed and a completed Share Form of Acceptance must be transmitted by facsimile to the Tender Agent on the same day that the shares are deposited to *Société Générale*. The original of this Share Form of Acceptance and the completed and signed tax form must be sent separately to the Tender Agent via overnight courier.

**THE OFFER HAS NOT BEEN SUBMITTED FOR CLEARANCE TO CONSOB (THE ITALIAN SECURITIES AND EXCHANGE COMMISSION). ACCORDINGLY, THE OFFER IS NOT BEING MADE AND WILL NOT BE MADE, DIRECTLY OR INDIRECTLY, IN ITALY, AND NO COPIES OF THIS DOCUMENT, THE OFFER TO PURCHASE OR ANY OTHER DOCUMENT RELATING TO THE OFFER HAVE BEEN OR WILL BE DISTRIBUTED IN ITALY, EXCEPT THAT, AS REQUIRED BY LAW, HOLDERS OF RECORD OR THOSE APPEARING ON A SECURITIES POSITION LISTING AS HOLDERS OF ORDINARY SHARES (INCLUDING, PURSUANT TO AN EXEMPTION AVAILABLE UNDER ITALIAN LAW, THE VERY LIMITED NUMBER OF SUCH HOLDERS LOCATED OR RESIDENT IN ITALY) WILL BE SENT COPIES OF THIS DOCUMENT AND OTHER DOCUMENTS RELATING TO THE OFFER. NEITHER THIS DOCUMENT NOR THE OFFER TO PURCHASE NOR ANY OTHER DOCUMENT RELATING TO THE OFFER MAY BE DISTRIBUTED TO ANY OTHER PERSON LOCATED OR RESIDENT IN ITALY FOR ANY REASON. NEITHER ANY SUCH DOCUMENT NOR ANY INFORMATION CONTAINED HEREIN OR THEREIN CONSTITUTES AN OFFER TO PURCHASE OR A SOLICITATION OF AN OFFER TO SELL OR AN ADVERTISEMENT OF AN OFFER TO PURCHASE ORDINARY SHARES IN ITALY WITHIN THE MEANING OF ARTICLE 1, PARAGRAPH 1, LETTER (V) OF ITALIAN LEGISLATIVE DECREE N. 58 OF FEBRUARY 24, 1998.**

**IN THE EVENT OF AN INCONSISTENCY BETWEEN THE TERMS AND CONDITIONS IN THIS SHARE FORM OF ACCEPTANCE AND THE OFFER DOCUMENTS, THE TERMS AND PROCEDURES IN THE OFFER DOCUMENT SHALL GOVERN.**

**NOTE: SIGNATURES MUST BE PROVIDED BELOW**  
**PLEASE READ THE ACCOMPANYING INSTRUCTIONS CAREFULLY**

Ladies and Gentlemen:

The undersigned hereby tenders to Purchaser the above-described Ordinary Shares pursuant to the Offer to Purchase, at a price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, on the terms and subject to the conditions set forth in the Offer to Purchase, receipt of which is hereby acknowledged, and this Share Form of Acceptance (which, together with the Offer to Purchase and the ADS Letter of Transmittal, as amended or supplemented from time to time, collectively constitute the “Offer”).

The undersigned understands that the acceptance for payment by Purchaser of Ordinary Shares tendered pursuant to one of the procedures described in the section “THE TENDER OFFER – Procedures for Tendering Ordinary Shares or ADSs” of the Offer to Purchase will constitute a binding agreement between the undersigned and Purchaser upon the terms and subject to the conditions of the Offer.

On the terms and subject to the conditions of the Offer (including, if the Offer is extended or amended, the terms and conditions of such extension or amendment) and to the extent permitted under applicable law, subject to, and effective upon, acceptance for payment and payment for the Ordinary Shares validly tendered herewith in accordance with the terms of the Offer, the undersigned hereby sells, assigns and transfers to, or upon the order of, Purchaser, all right, title and interest in and to all of the Ordinary Shares being tendered hereby and any and all cash dividends, distributions, rights, other Ordinary Shares, ADSs or other securities issued or issuable in respect of such Ordinary Shares on or after December 23, 2013 (collectively, “Distributions”). In addition, the undersigned irrevocably appoints The Bank of New York Mellon (the “Tender Agent”) as the true and lawful agent and attorney-in-fact of the undersigned with respect to such Ordinary Shares and any Distributions with full power of substitution (such power of attorney being deemed to be an irrevocable power coupled with an interest) to the fullest extent of the undersigned’s rights with respect to such Ordinary Shares and any Distributions to (a) deliver any Distributions, and to transfer ownership of Ordinary Shares and any Distributions on the account books maintained by DTC, together, in either such case, with all accompanying evidence of transfer and authenticity, to or upon the order of Purchaser, (b) present such Ordinary Shares and any Distributions for transfer on the books of Gentium and (c) receive all benefits and otherwise exercise all rights of beneficial ownership of such Ordinary Shares and any Distributions, all in accordance with the terms and subject to the conditions of the Offer and to the extent permitted under applicable law.

To the extent permitted under applicable law, the undersigned hereby irrevocably appoints each of the designees of Purchaser the attorneys-in-fact and proxies of the undersigned, each with full power of substitution, to the full extent of the undersigned’s rights with respect to the Ordinary Shares tendered hereby that have been accepted for payment and with respect to any Distributions. The designees of Purchaser will, with respect to the Ordinary Shares and any associated Distributions for which the appointment is effective, be empowered to exercise all voting and any other rights of the undersigned, as they, in their sole discretion, may deem proper. This proxy and power of attorney shall be irrevocable and coupled with an interest in the tendered Ordinary Shares. Such appointment is effective when, and only to the extent that, Purchaser accepts the Ordinary Shares tendered with this Share Form of Acceptance for payment pursuant to the Offer. Upon the effectiveness of such appointment, without further action, all prior powers of attorney, proxies and consents given by the undersigned with respect to such Ordinary Shares and any associated Distributions will be revoked and no subsequent powers of attorney, proxies, consents or revocations may be given (and, if given, will not be deemed effective). Purchaser reserves the right to require that, in order for Ordinary Shares to be deemed validly tendered, immediately upon Purchaser’s acceptance for payment of such Ordinary Shares, Purchaser must be able to exercise full voting, consent and other rights, to the extent permitted under applicable law, with respect to such Ordinary Shares and any associated Distributions, including voting at any meeting of shareholders concerning any matter.

The undersigned hereby represents and warrants that the undersigned has full power and authority to tender, sell, assign and transfer the Ordinary Shares and any Distributions tendered hereby and, when the same are accepted for payment by Purchaser, Purchaser will acquire good, marketable and unencumbered title thereto, free and clear of all liens, restrictions, charges and encumbrances and the same will not be subject to any adverse claim. The undersigned hereby represents and warrants that the undersigned is the registered owner of the Ordinary Shares, or the name of the undersigned appears on a security position listing maintained by Monte Titoli as owner of the Ordinary Shares. The undersigned will, upon request, execute and deliver any additional documents deemed by the Tender Agent or Purchaser to be necessary or desirable to complete the sale, assignment and transfer of the Ordinary Shares and any Distributions tendered hereby. In addition, the undersigned shall promptly remit and transfer to the Tender Agent for the account of Purchaser any and all Distributions in respect of the Ordinary Shares tendered hereby, accompanied by appropriate documentation of transfer and, pending such remittance or appropriate assurance thereof, Purchaser shall be entitled to all rights and privileges as owner of any such Distributions and may withhold the entire purchase price or deduct from the purchase price the amount or value thereof, as determined by Purchaser in its sole discretion.

It is understood that the undersigned will not receive payment for the Ordinary Shares unless and until the Ordinary Shares are accepted for payment or, in the case of Ordinary Shares held in book-entry form, ownership of Ordinary Shares is validly transferred on the account books maintained by Monte Titoli, and until the same are processed for payment by the Tender Agent. It is understood that the method of delivery of the Ordinary Shares and all other required documents is at the option and risk of the undersigned and that the risk of loss of such Ordinary Shares shall pass only after the Tender Agent has actually received the Ordinary Shares.

All authority conferred or agreed to be conferred pursuant to this Share Form of Acceptance shall not be affected by, and shall survive, the death or incapacity of the undersigned and any obligation of the undersigned hereunder shall be binding upon the heirs, executors, administrators, trustees in bankruptcy, personal representatives, successors and assigns of the undersigned. Except as stated in the Offer to Purchase, this tender is irrevocable.

Please issue the check for the purchase price in the name(s) of the owner(s) appearing under "Description of Ordinary Shares Tendered."

**IMPORTANT – SIGN HERE**  
**(U.S. Holders Please Also Complete Substitute Form W-9 Included Herein)**  
**(Non-U.S. Holders Please Obtain and Complete Form W-8BEN or Other Form W-8)**

(all holders must sign)

\_\_\_\_\_  
\_\_\_\_\_  
**(Signature(s) of Holder(s))**

Dated: \_\_\_\_\_, 20\_\_

Name(s): \_\_\_\_\_  
\_\_\_\_\_  
**(Please Print)**

Capacity (full title): \_\_\_\_\_  
\_\_\_\_\_

Address: \_\_\_\_\_  
\_\_\_\_\_  
**(Include Zip Code)**

Area Code and Telephone Number: \_\_\_\_\_

Tax Identification or  
Social Security No.: \_\_\_\_\_

**INSTRUCTIONS**  
**Forming Part of the Terms and Conditions of the Offer**

**1. Delivery of Share Form of Acceptance Confirmations of Share Deposit in Tender Agent's account at Société Générale.** This Share Form of Acceptance is to be used if tenders are to be made pursuant to the procedures for tender by book-entry transfer set forth in the section "THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs" of the Offer to Purchase. Confirmation of any book-entry transfer into the Tender Agent's account at *Société Générale* of Ordinary Shares tendered via Monte Tiotoli, as well as this Share Form of Acceptance properly completed and signed together with a completed and signed tax form duly executed must be received by the Tender Agent at one of its addresses set forth herein on or prior to the Expiration Date (as described in the section "THE TENDER OFFER – Terms of the Offer" of the Offer to Purchase).

**THE METHOD OF DELIVERY OF THE ORDINARY SHARES, THIS SHARE FORM OF ACCEPTANCE AND THE TAX DOCUMENT IS AT THE ELECTION AND RISK OF THE TENDERING HOLDER. DELIVERY OF ALL SUCH DOCUMENTS WILL BE DEEMED MADE AND RISK OF LOSS OF THE ORDINARY SHARES SHALL PASS ONLY WHEN ACTUALLY RECEIVED BY THE TENDER AGENT.**

No alternative, conditional or contingent tenders will be accepted and no fractional Ordinary Shares will be purchased. All tendering holders, by execution of this Share Form of Acceptance (or an originally signed facsimile thereof), waive any right to receive any notice of the acceptance of their Ordinary Shares for payment.

All questions as to validity, form, eligibility (including time of receipt) and acceptance for payment of any tender of Ordinary Shares hereunder will be determined by Purchaser, in its sole discretion (which may delegate power in whole or in part to the Tender Agent) and such determination shall be final and binding. Purchaser reserves the right to waive any irregularities or defects in the surrender of any Ordinary Shares. A surrender will not be deemed to have been made until all irregularities have been cured or waived. None of Purchaser, Parent or any of their respective affiliates or assigns, the Tender Agent, the Information Agent (as defined in the Offer to Purchase), or any other person will be under any duty to give notification of any defects or irregularities in tenders or incur any liability for failure to give any such notification. Purchaser's interpretation of the terms and conditions of the Offer (including this Share Form of Acceptance and the instructions hereto and any other documents related to the Offer) will be final and binding.

**2. Inadequate Space.** If the space provided herein is inadequate, the certificate numbers and/or the number of Ordinary Shares should be listed on a separate schedule attached hereto and separately signed on each page thereof in the same manner as this Share Form of Acceptance is signed.

**3. Requests for Assistance or Additional Copies.** Questions or requests for assistance may be directed to the Information Agent at its address and telephone number set forth below or to your broker or other financial intermediary. Additional copies of the Offer to Purchase, this Share Form of Acceptance and other tender offer materials may be obtained from the Information Agent as set forth below, subject to applicable law, and will be furnished at Purchaser's expense.

**4. Backup Withholding.** In order to avoid U.S. federal backup withholding with respect to cash received in exchange for Ordinary Shares pursuant to the Offer, a holder who is a U.S. person submitting Ordinary must (i) provide the Tender Agent with a properly completed Substitute Form W-9, included in this Share Form of Acceptance, or IRS Form W-9 and sign such form under penalties of perjury, (ii) provide the Tender Agent with a properly completed IRS Form W-8BEN or other Form W-8, and sign such form under penalties of perjury, or (iii) otherwise establish an exemption. IRS Form W-9, and W-8BEN and other Forms W-8 (if you are not a U.S. person) are available from the Tender Agent or from the Internal Revenue Service web site, at <http://www.irs.ustreas.gov>.



5. **Waiver of Conditions.** Subject to the terms and conditions of the Offer to Purchase and the applicable rules and regulations of the Securities and Exchange Commission, Purchaser reserves the right to waive in whole or in part at any time and from time to time in its sole discretion any of the conditions of the Offer prior to expiration of the Offer.

**IMPORTANT: THIS SHARE FORM OF ACCEPTANCE (OR AN ORIGINALLY SIGNED FACSIMILE THEREOF) TOGETHER WITH A COMPLETED AND SIGNED TAX FORM AND THE DEPOSIT OF THE SHARES IN THE TENDER AGENT'S ACCOUNT AT SOCIÉTÉ GÉNÉRALE MUST BE RECEIVED BY THE TENDER AGENT ON OR PRIOR TO THE EXPIRATION DATE.**

## IMPORTANT TAX INFORMATION

Under the U.S. federal income tax law, a holder who is a U.S. person whose tendered Ordinary Shares are accepted for purchase is required by law to (i) provide the Tender Agent with such holder's correct TIN on Substitute Form W-9 below, certify that such TIN is correct (or that such security holder is awaiting a TIN), and certify whether the holder is subject to backup withholding or (ii) otherwise establish a basis for exemption from backup withholding. If such holder of Ordinary Shares is an individual, the TIN is his or her social security number. If a holder of Ordinary Shares fails to provide a correct TIN to the Tender Agent, such person may be subject to a U.S.\$50.00 penalty imposed by the Internal Revenue Service. In addition, payments that are made to such holder of Ordinary Shares with respect to Ordinary Shares purchased pursuant to the Offer may be subject to backup withholding of 28%.

Certain holders of Ordinary Shares (including, among others, all corporations and certain foreign individuals) are not subject to these backup withholding and reporting requirements. In order for a foreign person to qualify for an exemption from backup withholding, that holder must generally submit a Form W-8BEN (or other applicable Form W-8), signed under penalties of perjury, attesting to that individual's exempt status. A Form W-8BEN can be obtained from the Tender Agent or the IRS website, at <http://www.irs.ustreas.gov>.

If backup withholding applies, the Tender Agent is required to withhold 28% of any payments made to the Ordinary Share or ADS holder or payee. Backup withholding is not an additional tax. Rather, the U.S. federal income tax liability of persons subject to backup withholding will be reduced by the amount of tax withheld. If withholding results in an overpayment of taxes, a refund may be obtained from the Internal Revenue Service by timely filing a claim for refund.

If backup withholding applies and "Applied For" is written in Part I of the Substitute Form W-9 and the holder of Ordinary Shares has completed the Certificate of Awaiting Taxpayer Identification Number, the Tender Agent will retain 28% of any payment of the purchase price for tendered Ordinary Shares during the 60-day period following the date of the Substitute Form W-9. If a holder's TIN is provided to the Tender Agent within 60 days of the date of the Substitute Form W-9, payment of such retained amounts will be made to such holder of Ordinary Shares. If a holder's TIN is not provided to the Tender Agent within such 60-day period, the Tender Agent will remit such retained amounts to the Internal Revenue Service as backup withholding and shall withhold 28% of any payment of the purchase price for the tendered Ordinary Shares made to such holder thereafter unless such holder of Ordinary Shares furnishes a TIN to the Tender Agent prior to such payment.

### **Purpose of Substitute Form W-9**

To prevent backup withholding on payments made to a holder whose tendered Ordinary Shares are accepted for purchase (for holders other than foreign persons who provide an appropriate Form W-8BEN), the holder should complete and sign the Substitute Form W-9 included in this Share Form of Acceptance and provide the holder's correct TIN and certify, under penalties of perjury, that the TIN provided on such Form is correct (or that such holder is awaiting a TIN) and that (a) such holder of Ordinary Shares is exempt from backup withholding, (b) such holder of Ordinary Shares has not been notified by the Internal Revenue Service that such holder is subject to backup withholding as a result of failure to report all interest or dividends or (c) the Internal Revenue Service has notified the holder that the holder is no longer subject to backup withholding. The holder must sign and date the Substitute Form W-9 where indicated, certifying that the information on such Form is correct.

### **What Number to Give the Tender Agent on the Substitute Form W-9**

The holder of Ordinary Shares is required to give the Tender Agent the social security number or employer identification number of the record owner of the Ordinary Shares. If the Ordinary Shares are in more than one name or are not in the name of their actual holder, consult the enclosed Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9 for additional guidance on which number to report.

**TO BE COMPLETED BY ALL TENDERING U.S. HOLDERS**

(See Instruction 9)

**PAYER: The Bank of New York Mellon**

<p><b>SUBSTITUTE</b></p> <p><b>Form W-9</b></p> <p><b>Request for Taxpayer Identification Number (TIN) and Certification</b></p>	Name: _____	
	Address: _____	
	Check appropriate box:	
	Individual/Sole Proprietor <input type="checkbox"/>	Limited Liability Company. Enter the tax classification:
	S Corporation <input type="checkbox"/>	C corporation <input type="checkbox"/>
C Corporation <input type="checkbox"/>	S corporation <input type="checkbox"/>	
Trust/Estate <input type="checkbox"/>	Partnership <input type="checkbox"/>	
Partnership <input type="checkbox"/>	Other (specify) <input type="checkbox"/>	
<b>Part I.</b> Please provide your taxpayer identification number in the space at right. If awaiting TIN, write "Applied For" in space at right and complete the Certificate of Awaiting Taxpayer Identification Number below.	SSN:	
	Or	
	EIN:	
<b>Part II.</b> For Payees exempt from backup withholding, see the enclosed "Guidelines for Certification of Taxpayer Identification Number on Substitute Form W-9" and complete as instructed therein.		
<b>Part III. CERTIFICATION</b>		
Under penalties of perjury, I certify that:		
(1) The number shown on this form is my correct Taxpayer Identification Number (or, as indicated, I am waiting for a number to be issued to me);		
(2) I am not subject to backup withholding because: (a) I am exempt from backup withholding, or (b) I have not been notified by the IRS that I am subject to backup withholding as a result of a failure to report all interests or dividends, or (c) the IRS has notified me that I am no longer subject to backup withholding; and		
(3) I am a U.S. citizen or other U.S. person (including a U.S. resident alien).		
Certification Instructions – You must cross out item (2) above if you have been notified by the IRS that you are subject to backup withholding because of underreporting of interest or dividends on your tax return. However, if after being notified by the IRS that you were subject to backup withholding you received another notification from the IRS that you are no longer subject to backup withholding, do not cross out item (2).		
Signature of U.S. person: _____		Date: _____

**YOU MUST COMPLETE THE FOLLOWING CERTIFICATE IF YOU WROTE "APPLIED FOR" IN PART I OF THIS SUBSTITUTE FORM W-9**

**CERTIFICATE OF AWAITING TAXPAYER IDENTIFICATION NUMBER**

I certify under penalties of perjury that a taxpayer identification number has not been issued to me, and either (a) I have mailed or delivered an application to receive a taxpayer identification number to the appropriate Internal Revenue Service Center or Social Security Administration Office or (b) I intend to mail or deliver an application in the near future. I understand that, notwithstanding the information I provided in Part III of the Substitute Form W-9 (and the fact that I have completed this Certificate of Awaiting Taxpayer Identification Number), all reportable payments made to me hereafter will be subject to a 28 percent backup withholding tax until I provide a properly certified taxpayer identification number.

\_\_\_\_\_  
Signature

\_\_\_\_\_  
Date

**NOTE: FAILURE TO COMPLETE AND RETURN THIS FORM MAY RESULT IN BACKUP WITHHOLDING OF 28% OF ANY PAYMENTS MADE TO YOU PURSUANT TO THIS OFFER. PLEASE REVIEW THE ENCLOSED GUIDELINES FOR CERTIFICATION OF TAXPAYER IDENTIFICATION NUMBER ON SUBSTITUTE FORM W-9 FOR ADDITIONAL DETAILS.**

***The Tender Agent for the Offer is:  
The Bank of New York Mellon***

*By Registered, Certified or Express Mail:*  
The Bank of New York Mellon  
Voluntary Corporate Actions  
P.O. Box 3031  
Providence, Rhode Island 04940-3031  
United States of America

*By Overnight Courier:*  
The Bank of New York Mellon  
Voluntary Corporate Actions – Suite V  
250 Royall Street  
Canton, Massachusetts 02021  
United States of America

***The Information Agent for the Offer is:***

***MACKENZIE  
PARTNERS, INC***

105 Madison Avenue  
New York, New York 10016  
(212) 929-5500 (Call Collect)  
or  
(800) 322-2885 (Toll Free)  
Email: [tenderoffer@mackenziepartners.com](mailto:tenderoffer@mackenziepartners.com)

December 23, 2013

VOLUNTARY CORPORATE ACTIONS COY: GENB

*This announcement is neither an offer to purchase nor a solicitation of an offer to sell Ordinary Shares or ADSs (each as defined below). The Offer (as defined below) is made solely by the Offer to Purchase (as defined below) and the related Share Form of Acceptance and ADS Letter of Transmittal (each as defined below) and any amendments or supplements to the Offer to Purchase, Share Form of Acceptance or ADS Letter of Transmittal and, other than as described below, is being made to all holders of Ordinary Shares and ADSs. The Offer is not being made to (and tenders will not be accepted from or on behalf of) holders of Ordinary Shares or ADSs in any jurisdiction in which the making or acceptance of the Offer would not be in compliance with the laws of such jurisdiction or any administrative or judicial action pursuant thereto. However, Purchaser (as defined below) may, in its discretion, take such action as it may deem necessary to make the Offer in any jurisdiction and extend the Offer to holders of Ordinary Shares and ADSs in such jurisdiction.*

**Notice of Offer to Purchase for Cash**

**All Outstanding Ordinary Shares and American Depositary Shares**

**(each American Depositary Share representing one Ordinary Share)**

of

**GENTIUM S.p.A.**

at

**\$57.00 per Ordinary Share and**

**per American Depositary Share**

by

**JAZZ PHARMACEUTICALS ITALY S.r.l.**

a wholly-owned subsidiary of

**JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY**

Jazz Pharmaceuticals Italy S.r.l., an Italian *società a responsabilità limitata* (“Purchaser”) and a wholly-owned subsidiary of Jazz Pharmaceuticals Public Limited Company, a public limited company formed under the laws of Ireland (“Parent”), is offering to purchase all outstanding shares of ordinary stock, no par value per share (“Ordinary Shares”) of Gentium S.p.A., a *società per azioni* incorporated in Italy (the “Company”) and all outstanding American Depositary Shares (“ADSs”), each representing one Ordinary Share and evidenced by an American Depositary Receipt (“ADR”) at a purchase price of \$57.00 per Ordinary Share and per ADS (without duplication for Ordinary Shares underlying ADSs), net to the seller in cash, without interest thereon and less any required withholding taxes, upon the terms and subject to the conditions set forth in the Offer to Purchase, dated December 23, 2013 (the “Offer to Purchase”) and in the related Share Form of Acceptance (the “Share Form of Acceptance”) and ADS Letter of Transmittal (the “ADS Letter of Transmittal” and, together with the Share Form of Acceptance and Offer to Purchase as amended or supplemented from time to time, the “Offer”). Tendering holders who have Ordinary Shares registered in an account held at Monte Titoli S.p.A. (“Monte Titoli”) or ADSs registered on the books of The Bank of New York Mellon (the “ADS Depository”) will not be obligated to pay brokerage fees or commissions or, except as set forth in Instruction 6 of each of the Share Form of Acceptance and ADS Letter of Transmittal, transfer taxes on the purchase of Ordinary Shares or ADSs by Purchaser pursuant to the Offer. Holders who hold their Ordinary Shares or ADSs through a broker or bank should consult with such institution as to whether it charges any service fees. Unless Purchaser extends the expiration date of the Offer in accordance with the terms of the Tender Offer Agreement (as defined below) and applicable law (as described below), the Offer and withdrawal rights will expire at 12:00 midnight, New York City time, on the evening of January 22, 2014 for all Ordinary Shares and ADSs (such initial or extended expiration date in accordance with the terms of the Tender Offer Agreement and applicable law, the “Expiration Date”). Purchaser will pay the fees and expenses incurred in connection with the Offer of the Tender Agent (as defined below) and MacKenzie Partners, Inc., the information agent for the Offer (the “Information Agent”). Purchaser is offering to purchase all Ordinary Shares and ADSs to acquire control of, and the entire equity interest in, the Company.

**THE OFFER AND WITHDRAWAL RIGHTS WILL EXPIRE AT 12:00 MIDNIGHT, NEW YORK CITY TIME, ON THE EVENING OF JANUARY 22, 2014, UNLESS THE OFFER IS EXTENDED. ORDINARY SHARES AND ADSs TENDERED PURSUANT TO THIS OFFER MAY BE WITHDRAWN AT ANY TIME PRIOR TO THE EXPIRATION OF THE OFFER, BUT NOT DURING ANY SUBSEQUENT OFFERING PERIOD.**

**THE OFFER HAS NOT BEEN SUBMITTED FOR CLEARANCE TO CONSOB (THE ITALIAN SECURITIES AND EXCHANGE COMMISSION). ACCORDINGLY, THE OFFER IS NOT BEING MADE AND WILL NOT BE MADE, DIRECTLY OR INDIRECTLY, IN ITALY AND NO COPIES OF THIS DOCUMENT, THE OFFER TO PURCHASE OR ANY OTHER DOCUMENT RELATING TO THE OFFER HAVE BEEN OR WILL BE DISTRIBUTED IN ITALY, EXCEPT THAT, AS REQUIRED BY LAW, HOLDERS OF RECORD OR THOSE APPEARING ON A SECURITIES POSITION LISTING AS HOLDERS OF ADSs OR ORDINARY SHARES (INCLUDING, PURSUANT TO AN EXEMPTION AVAILABLE UNDER ITALIAN LAW, THE VERY LIMITED NUMBER OF SUCH HOLDERS LOCATED OR RESIDENT IN ITALY) WILL BE SENT COPIES OF CERTAIN DOCUMENTS RELATING TO THE OFFER. NEITHER THIS DOCUMENT NOR THE OFFER TO PURCHASE NOR ANY OTHER DOCUMENT RELATING TO THE OFFER MAY BE DISTRIBUTED TO ANY OTHER PERSON LOCATED OR RESIDENT IN ITALY FOR ANY REASON. NEITHER ANY SUCH DOCUMENT NOR ANY INFORMATION CONTAINED HEREIN OR THEREIN CONSTITUTES AN OFFER TO PURCHASE OR A SOLICITATION OF AN OFFER TO SELL OR AN ADVERTISEMENT OF AN OFFER TO PURCHASE ORDINARY SHARES OR ADSs IN ITALY WITHIN THE MEANING OF ARTICLE I, PARAGRAPH 1, LETTER (V) OF ITALIAN LEGISLATIVE DECREE N. 58 OF FEBRUARY 24, 1998.**

Following consummation of the Offer, Purchaser intends to cause the Company to seek to delist the ADSs from the Nasdaq Global Select Market and to cause the Company to terminate the existing deposit agreement between the Company and the ADS Depositary, dated as of June 15, 2005 (the "Deposit Agreement"), under which the ADSs were issued. If the Deposit Agreement is terminated, holders of ADSs will be entitled to receive one Ordinary Share in exchange for each ADS held by such holders upon surrender by them of their ADSs for at least one year following such termination. At any time after the expiration of one year from the date of termination, the ADS Depositary may sell the Ordinary Shares underlying ADSs that have not been surrendered and hold uninvested the proceeds on behalf of holders. Purchaser intends, to the extent possible following the consummation of the Offer, to seek to cause the Company to deregister the Ordinary Shares and the ADSs under the Securities Exchange Act of 1934, as amended (the "Exchange Act"), so that the Company will no longer be obligated to file any reports with the Securities and Exchange Commission (the "Commission") or otherwise be required to comply with the Commission's rules relating to publicly held companies. Holders will not have appraisal rights in connection with the Offer. Purchaser's intentions and the effects of the Offer are more fully described in the Offer to Purchase.

The Offer is being made pursuant to a Tender Offer Agreement, dated as of December 19, 2013 (as it may be amended from time to time, the "Tender Offer Agreement"), by and among Parent, Purchaser and the Company.

**The Company's Board of Directors (the "Company Board") has determined that the Tender Offer Agreement and the transactions contemplated thereby are fair to and in the best interests of the Company's shareholders and other stakeholders of the Company. The Company Board has approved and deemed advisable the Tender Offer Agreement and the transactions contemplated thereby and has recommended that the holders of Ordinary Shares and ADSs accept the Offer and tender their Ordinary Shares and ADSs to Purchaser pursuant to the Offer.**

Consummation of the Offer is subject to various conditions set forth in the Tender Offer Agreement, including, but not limited to the valid tender (excluding ADSs tendered pursuant to guaranteed delivery procedures that are not yet delivered in settlement or satisfaction of such guarantee), without proper withdrawal, of a number of Ordinary Shares and ADSs that, together with any Ordinary Shares and ADSs owned by Parent or

Purchaser or any of their subsidiaries, represents at least 66 2/3% of the sum of (A) the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the time that Purchaser first accepts any Ordinary Shares and ADSs for payment pursuant to the Offer (the "Acceptance Time"), plus (B) the aggregate number of Ordinary Shares and ADSs issuable upon the conversion, exchange, settlement or exercise, as applicable, of all options and other rights to acquire, or securities convertible into or exchangeable for, Ordinary Shares and ADSs that are outstanding immediately prior to the Acceptance Time in all cases without duplication for Ordinary Shares underlying the ADSs, provided that Parent may, in its sole discretion, elect to reduce such minimum tender condition to a majority of the aggregate number of outstanding Ordinary Shares and ADSs immediately prior to the Acceptance Time (without duplication for Ordinary Shares underlying ADSs). Subject to the terms of the Tender Offer Agreement, Purchaser and Parent reserve the absolute right in its sole discretion to waive any of the specified conditions of the Offer in the case of any Ordinary Shares and ADSs tendered other than the condition set forth above. See the section in the Offer to Purchase entitled "THE TENDER OFFER – Certain Conditions to the Offer" for a more complete description of the conditions to the Offer.

For purposes of the Offer, Purchaser will be deemed to have accepted for payment and thereby purchased Ordinary Shares and ADSs validly tendered and not properly withdrawn if and when Purchaser gives notice to The Bank of New York Mellon (the "Tender Agent") of its acceptance for payment of such Ordinary Shares and ADSs pursuant to the Offer. Payment for Ordinary Shares and ADSs accepted for payment pursuant to the Offer will be made by deposit of the purchase price therefor with the Tender Agent, which will act as agent for the tendering holders for purposes of receiving payments from Purchaser and transmitting such payments to the tendering holders. **Under no circumstances will Purchaser pay interest on the purchase price for Ordinary Shares or ADSs, regardless of any extension of the Offer or any delay in payment for Ordinary Shares or ADSs.**

In all cases, (a) payment for Ordinary Shares tendered and accepted for payment pursuant to the Offer will be made only after timely receipt by the Tender Agent of: (i) a timely book-entry transfer of such Ordinary Shares into the Monte Titoli account of the Tender Agent's Custodian (as defined in the Offer to Purchase) pursuant to the procedures set forth in "THE TENDER OFFER – Procedure for Tendering Ordinary Shares and ADSs" in the Offer to Purchase; (ii) a Share Form of Acceptance (or a manually signed facsimile thereof) properly completed and duly executed; and (iii) any other documents required by the Share Form of Acceptance and (b) payment for ADSs tendered and accepted for payment pursuant to the Offer will be made only after timely receipt by the Tender Agent of: (i) certificates evidencing such ADSs representing such ADSs or a timely Book-Entry confirmation (as defined in the Offer to Purchase) of such ADSs into the Tender Agent's account at The Depository Trust Company ("DTC") pursuant to the procedures set forth in the section entitled "THE TENDER OFFER – Procedure for Tendering Ordinary Shares and ADSs" in the Offer to Purchase; (ii) an ADS Letter of Transmittal (or a manually signed facsimile thereof), properly completed and duly executed, with any required signature guarantees (or, in the case of a book-entry transfer, an Agent's Message (as defined in the section entitled "THE TENDER OFFER – Procedure for Tendering Ordinary Shares and ADSs" in the Offer to Purchase) in lieu of the ADS Letter of Transmittal); and (iii) any other documents required by the ADS Letter of Transmittal.

The Tender Offer Agreement and the U.S. securities laws permit Purchaser to extend the Offer and thereby delay acceptance for payment of, and the payment for, any Ordinary Shares or ADSs tendered in the Offer. Under the Tender Offer Agreement, Purchaser is required to extend the Offer for successive periods of up to 20 business days per extension if any condition to the Offer has not been satisfied or waived as of the scheduled expiration date of the Offer. In addition, Purchaser is also required to extend the Offer from time to time for any period that is required by any rule, regulation, interpretation or position of the Commission, its staff or the Nasdaq Stock Market that is applicable to the Offer. Purchaser is not required to extend the Offer beyond April 19, 2014, unless either Parent or the Company elects to extend this date to September 19, 2014, in the event that certain specified conditions are the only conditions that are not satisfied or waived on April 19, 2014.

During any extension of the Offer (as opposed to a subsequent offering period discussed below), all Ordinary Shares and ADSs previously tendered and not withdrawn will remain subject to the Offer and subject to withdrawal rights. Pursuant to the Tender Offer Agreement and Rule 14d-11 under the Exchange Act, Purchaser

will, and Purchaser reserves the right to, provide a subsequent offering period of three to 20 business days in length following the acceptance of tendered Ordinary Shares and ADSs upon expiration of the Offer on the Expiration Date (the “Subsequent Offering Period”). The Subsequent Offering Period will be an additional period of time, beginning no later than 9:00 a.m., New York City time, on the next business day following the expiration of the Offer, during which holders may tender Ordinary Shares and ADSs not tendered in the Offer. The Subsequent Offering Period is not an extension of the Offer, which already will have been completed. See the section entitled “THE TENDER OFFER – Terms of the Offer” in the Offer to Purchase.

Any extension, termination, waiver or amendment of the Offer will be followed, as promptly as practicable, with a public announcement to be issued no later than 9:00 a.m., New York City time, on the next business day after the previously scheduled Expiration Date. Subject to the requirements of the Exchange Act and other applicable law, and without limiting the manner in which Parent or Purchaser may choose to make any public announcement, Parent and Purchaser assume no obligation to publish, advertise or otherwise communicate any public announcement.

Tenders of Ordinary Shares and ADSs pursuant to the Offer are irrevocable, except that Ordinary Shares and ADSs tendered pursuant to the Offer may be withdrawn at any time on or prior to the Expiration Date and, unless theretofore accepted for payment by Purchaser pursuant to the Offer, may also be withdrawn at any time after March 24, 2014. For a withdrawal of Ordinary Shares or ADSs to be effective, a signed notice of withdrawal must be timely received by the Tender Agent at one of its addresses set forth on the back cover of the Offer to Purchase. Any notice of withdrawal must specify the name of the person having tendered Ordinary Shares or ADSs to be withdrawn, the number of Ordinary Shares or ADSs to be withdrawn and the name of the record holder of Ordinary Shares or ADRs representing ADSs to be withdrawn, if different from that of the person who tendered such Ordinary Shares or ADSs. The signature(s) on the notice of withdrawal must be medallion guaranteed by an Eligible Institution (as defined in the section entitled “THE TENDER OFFER – Procedure for Tendering Ordinary Shares and ADSs” in the Offer to Purchase), if the original letter of transmittal required a medallion signature guarantee in accordance with the Offer. If Ordinary Shares or ADSs have been tendered pursuant to the procedures for book-entry transfer as set forth in the section entitled “THE TENDER OFFER – Procedure for Tendering Ordinary Shares and ADSs” in the Offer to Purchase, any notice of withdrawal must specify the name and number of the account at Monte Titoli (with respect to Ordinary Shares) and DTC (with respect to ADSs) to be credited with the withdrawn Ordinary Shares and ADSs. If certificates evidencing ADRs representing ADSs to be withdrawn have been delivered or otherwise identified to the Tender Agent, the name of the registered owner and the serial numbers shown on such certificates or ADRs must also be furnished to the Tender Agent as aforesaid prior to the physical release of such certificates. All questions as to the form and validity (including time of receipt) of any notice of withdrawal will be determined by Purchaser, in its sole discretion, which determination will be final and binding. No withdrawal of Ordinary Shares or ADSs will be deemed to have been properly made until all defects and irregularities have been cured or waived. None of the Parent or Purchaser, any of their respective affiliates or assigns, the Tender Agent, the Information Agent or any other person will be under any duty to give notification of any defects or irregularities in any notice of withdrawal or incur any liability for failure to give such notification. Withdrawals of tenders of Ordinary Shares or ADSs may not be rescinded, and any Ordinary Shares or ADSs properly withdrawn will be deemed not to have been validly tendered for purposes of the Offer. However, withdrawn Ordinary Shares and ADSs may be re-tendered by following one of the procedures for tendering Ordinary Shares and ADSs described in the section entitled “THE TENDER OFFER – Procedures for Tendering Ordinary Shares and ADSs” in the Offer to Purchase at any time prior to the Expiration Date.

The exchange of Ordinary Shares or ADSs for cash pursuant to the Offer generally will be a taxable transaction for U.S. federal income tax purposes, and a U.S. holder who receives cash for Ordinary Shares or ADSs pursuant to the Offer will recognize gain or loss, if any, equal to the difference between the amount of cash received and such U.S. holder’s adjusted tax basis in Ordinary Shares or ADSs. Such gain or loss will be capital gain or loss, and will be long-term capital gain or loss if such U.S. holder’s holding period for Ordinary Shares or ADSs is more than one year at the time of the exchange of such U.S. holder’s Ordinary Shares or ADSs for cash. Long-term capital gains recognized by an individual U.S. holder generally are subject to tax at a lower rate than short-term capital gains or ordinary income. There are limitations on the deductibility of capital losses. For a



more complete description of certain U.S. federal income tax consequences of the Offer, see the section entitled “THE TENDER OFFER – Certain Tax Consequences of the Offer” in the Offer to Purchase.

The information required to be disclosed by paragraph (d)(1) of Rule 14d-6 of the General Rules and Regulations under the Exchange Act is contained in the Offer to Purchase and is incorporated herein by reference.

The Company has provided Purchaser with the Company’s list of holders of Ordinary Shares, and the ADS Depositary has provided Purchaser with a list of ADR holders and a security position listing for the purpose of disseminating the Offer to holders of Ordinary Shares and ADSs. The Offer to Purchase, the Share Form of Acceptance and the ADS Letter of Transmittal will be mailed to record holders of Ordinary Shares and ADSs (including, pursuant to an exemption available under Italian law, the very limited number of such holders located or resident in Italy) and will be furnished to brokers, dealers, commercial banks, trust companies and similar persons whose names, or the names of whose nominees, appear on the holder list or, if applicable, who are listed as participants in a clearing agency’s security position listing, for subsequent transmittal to beneficial owners of ADSs to the extent permitted by applicable law.

**The Offer to Purchase and the related Share Form of Acceptance and ADS Letter of Transmittal contain important information that should be read carefully and in their entirety before any decision is made with respect to the Offer.**

Questions and requests for assistance may be directed to the Information Agent at the addresses and telephone numbers set forth below. Requests for copies of the Offer to Purchase, Share Form of Acceptance, ADS Letter of Transmittal and other Offer documents may be directed to the Information Agent at its address and telephone number set forth below and copies will be furnished at Parent’s expense. Parent will not pay any fees or commissions to any broker or dealer or other person (other than to the Tender Agent or the Information Agent) for soliciting tenders of Ordinary Shares and ADSs in the Offer.

*The Information Agent for the Offer is:*

**MacKenzie Partners, Inc.**  
105 Madison Avenue  
New York, New York 10016  
(212) 929-5500 (Call Collect)  
1 (800) 322-2885 (Call Toll-Free)  
Email: tenderoffer@mackenziepartners.com

December 23, 2013

BARCLAYS  
745 Seventh Avenue  
New York, New York 10019

**PERSONAL AND CONFIDENTIAL**

December 19, 2013

Jazz Pharmaceuticals Public Limited Company  
45 Fitzwilliam Square  
Dublin 2, Ireland

Attention: Kate Falberg

**Project Genesis  
Commitment Letter**

Ladies and Gentlemen:

You have advised Barclays Bank PLC (the "Commitment Party," "Barclays," "we" or "us"), that Jazz Pharmaceuticals Public Limited Company (the "Buyer" or "you") intends to acquire (the "Acquisition") all of the ordinary stock and American Depositary Shares representing ordinary stock (the "Company Stock") of a company previously identified to us as "Genesis" (the "Company") in a cash tender offer (the "Tender Offer") pursuant to a Tender Offer Agreement to be entered into among you, a newly-formed wholly-owned subsidiary of yours and the Company (the definitive documentation for the Acquisition, together with the schedules and exhibits thereto, the "Acquisition Agreement") and to consummate certain transactions described therein (as described in Exhibit A and as otherwise contemplated by this Commitment Letter and the Fee Letter (each as defined below), the "Transactions"), in each case on the terms and subject to the conditions set forth in this Commitment Letter and Exhibits A and B (collectively, the "Commitment Letter").

You have also advised us that the total cost of the Acquisition (and related fees, commissions and expenses (collectively, "Transaction Costs")) will be financed with (i) a \$500 million senior secured incremental term loan facility (the "Incremental Term Facility") issued pursuant to the Credit Agreement dated June 12, 2012, as amended, by and among the Buyer, Jazz Pharmaceuticals, Inc., a Delaware corporation, Jazz Pharmaceuticals Ireland Limited, a company organized under the laws of Ireland, Jazz Financing I Limited, a company organized under the laws of Ireland, each of the lenders party thereto and Barclays Bank PLC, as administrative agent (the "Existing Credit Agreement"), having the terms set forth in Exhibit A and (ii) a combination of revolving loans drawn under the Existing Credit Agreement and cash on hand of the Buyer and/or its subsidiaries. Capitalized terms used in this Commitment Letter but not otherwise defined herein shall have the meanings set forth in the Existing Credit Agreement.

1. Commitments and Arranger Roles

You hereby appoint Barclays to act, and Barclays hereby agrees to act, as sole lead arranger, sole bookrunner and sole syndication agent (in such capacities, the "Arranger"), for the Incremental Term Facility, subject to the terms and conditions of this Commitment Letter and the Fee Letter. The Arranger will have the rights and authority customarily given to financial institutions in such roles. The Commitment Party is pleased to advise you of its commitment to provide to the Borrower 100% of the Incremental Term Facility on the terms and subject only to the conditions set forth in this Commitment Letter and the Fee Letter.

December 19, 2013

**Jazz Pharmaceuticals Public Limited Company**

Our fees for services related to the Incremental Term Facility are set forth in a separate fee letter (the "Fee Letter") between you and us entered into on the date hereof. As consideration for the execution and delivery of this Commitment Letter by us, you agree to pay the fees and expenses set forth in Exhibit A and in the Fee Letter as and when payable in accordance with the terms hereof and thereof. You agree that no other titles will be awarded in connection with the Incremental Term Facility unless you and we shall so agree.

**2. Conditions Precedent**

Our commitments hereunder and our agreements to perform the services described herein are subject to the following conditions: (i) the conditions set forth in Exhibit A under the heading "Conditions Precedent to Borrowing" and (ii) the conditions set forth in Exhibit B (collectively, the "Funding Conditions"); it being understood that there are no conditions (implied or otherwise) to each of the commitments hereunder (including compliance with the terms of the Commitment Letter, the Fee Letter and the Loan Documents) other than the Funding Conditions that are expressly stated to be conditions to the initial funding under the Incremental Term Facility (as such term is defined in Exhibit A) on the Closing Date (as such term is defined in Exhibit A).

Notwithstanding anything in this Commitment Letter, the Fee Letter, the Loan Documents or any other letter agreement between you and us concerning the financing of the Transactions to the contrary, the terms of the incremental amendment effecting the Incremental Term Facility (the "Incremental Amendment") will be in a form such that they do not impair the availability of the Incremental Term Facility on the Closing Date if the Funding Conditions are satisfied (it being understood that no further action shall be required with respect to the creation or perfection of any security interest in the Collateral on or prior to the Closing Date). This paragraph shall be referred to as the "Conditions Limitation Provision."

**3. Syndication**

The Arranger intends and reserves the right to syndicate the Incremental Term Facility to a group of Lenders (as such term is defined in Exhibit A) in consultation with you; provided that, unless otherwise agreed by you in writing, (i) no syndication will occur prior to the public announcement of the Acquisition by you, (ii) the Commitment Party will not be relieved, released or novated from its obligations hereunder (including its obligation to fund the Incremental Term Facility on the Closing Date), (iii) no assignment prior to the Closing Date will reduce or release the Commitment Party's obligation to fund its commitment in the event any assignee shall fail to do so on the Closing Date, and (iv) the Commitment Party shall retain exclusive control over all rights and obligations with respect to its commitment, including all rights with respect to consents, modifications, waivers and amendments, until the Closing Date has occurred.

The Arranger will lead the syndication, including determining the timing of all offers to prospective Lenders, any title of agent or similar designations or roles awarded to any Lender and the acceptance of commitments, the amounts offered and the compensation provided to each Lender from the amounts to be paid to the Arranger pursuant to the terms of this Commitment Letter and the Fee Letter and will, in consultation with you, determine the final commitment allocations. You agree to use commercially reasonable efforts to ensure that the Arranger's syndication efforts benefit from the existing lending and investment banking relationships of you and your subsidiaries and, to the extent appropriate, the Company and its subsidiaries until the date that is the earliest of (a) the termination by the Arranger of syndication of the Incremental Term Facility, (b) 60 days following the Closing Date and (c) the "successful syndication" of the Incremental Term Facility (as defined in the Fee Letter) (such earliest date, the "Syndication Date"). Prior to the Syndication Date, you will not, and agree to use commercially

December 19, 2013

**Jazz Pharmaceuticals Public Limited Company**

reasonable efforts to obtain contractual undertakings from the Company that it will not, without the prior written consent of the Arranger, syndicate or issue, attempt to syndicate or issue or announce the syndication or issuance of, any competing debt facility or any competing debt security of the Company or you or any of your or its respective subsidiaries or affiliates, including any renewal or refinancing of any existing debt facility or debt security, in each case without the prior written consent of the Arranger (other than the Incremental Term Facility and purchase money and capital lease financings); provided that no such contractual undertakings shall be required to be obtained from the Company prior to the execution of the Acquisition Agreement.

You agree to, and agree to use commercially reasonable efforts to obtain contractual undertakings from the Company to, cooperate with, and provide information reasonably required by, the Arranger in connection with all syndication efforts, including: (i) your assistance in the preparation as soon as practicable after execution of the Acquisition Agreement, a customary information memorandum and other customary presentation materials (collectively, "Marketing Materials") reasonably acceptable in form and content to the Arranger regarding the business, operations, financial projections and prospects of you and the Company (including the financial information described in Exhibit B) including without limitation the delivery of all information relating to the Transactions prepared by or on behalf of you or the Company that the Arranger deems reasonably necessary to complete the syndication of the Incremental Term Facility; (ii) using your commercially reasonable efforts to obtain from Moody's Investor Service, Inc. ("Moody's") and Standard & Poor's Ratings Services, a Standard & Poor's Financial Services LLC business ("S&P"), prior to the launch of the general syndication, a corporate family rating and a credit rating for the Incremental Term Facility (it being understood that no such rating shall be required to be obtained); (iii) your arranging for direct communications with prospective Lenders in connection with the syndication of the Incremental Term Facility (including without limitation direct contact between your appropriate senior management, representatives and advisors (and, from and after execution of the Acquisition Agreement, using commercially reasonable efforts to cause direct contact with appropriate senior management, representatives and advisors of the Company) and participation of such persons in such meetings, in each case upon reasonable prior notice and at places and times and frequency to be mutually agreed); and (iv) your hosting (including any preparations with respect thereto) with the Arranger at places and times to be mutually agreed a reasonable number of meetings to be mutually agreed with prospective Lenders. You agree that the Arranger has the right to place advertisements in financial and other newspapers at its own expense describing its services to you; provided that no such advertisement shall be placed prior to the public announcement of the Acquisition by you and the Arranger shall submit a copy of any such advertisements to the Buyer for its prior approval, which approval shall not be unreasonably withheld, conditioned or delayed. Notwithstanding anything to the contrary contained in this Commitment Letter or the Fee Letter, the obtaining of the ratings referred above shall not constitute a condition to the commitment hereunder or the funding of the Incremental Term Facility on the Closing Date.

You will be solely responsible for the contents of the Marketing Materials and all other information, documentation or other materials delivered to us in connection therewith and you acknowledge that we will be using and relying upon such information without independent verification thereof.

You understand that certain prospective Lenders (such Lenders, "Public Lenders") may have personnel that do not wish to receive MNPI (as defined below). At the Arranger's request, you agree to assist in the preparation of an additional version of the Marketing Materials that does not contain material non-public information concerning you, the Company or your or its respective subsidiaries or affiliates or your or its respective securities within the meaning of United States federal and state securities laws (collectively, "MNPI") which is suitable to make available to Public Lenders. You acknowledge and agree that the following documents may be distributed to Public Lenders (except to the extent you notify us to the contrary and provided that you shall have been given a reasonable opportunity to review such documents

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and comply with the U.S. Securities and Exchange Commission disclosure requirements): (a) drafts and final versions of the Incremental Amendment and the other Loan Documents; (b) administrative materials prepared by the Arranger for prospective Lenders (including without limitation a lender meeting invitation, allocations and funding and closing memoranda); and (c) term sheets and notification of changes in the terms and conditions of the Incremental Term Facility. Before distribution of any Marketing Materials in connection with the syndication of the Incremental Term Facility (i) to prospective Lenders that are not Public Lenders, you will provide us with a customary letter authorizing the dissemination of such materials and (ii) to prospective Public Lenders, you will provide us with a customary letter authorizing the dissemination of information that does not contain MNPI (the "Public Information Materials") to Public Lenders and confirming the absence of MNPI therein; provided that any such authorization letters shall be reasonably satisfactory to you. In addition, at the Arranger's request, you will identify Public Information Materials by marking the same as "PUBLIC" (it being understood that you shall not be under any obligation to mark any materials as "PUBLIC").

Without limiting your obligations to assist with the syndication efforts as set forth herein, it is understood that the Commitment Party's commitments hereunder are not conditioned upon the syndication of, or receipt of commitments in respect of, the Incremental Term Facility, and in no event shall the commencement or successful completion of syndication of the Incremental Term Facility constitute a condition to the availability of the Incremental Term Facility on the Closing Date. You hereby acknowledge and agree that the Arranger, in such capacity, will have no responsibility other than to arrange the syndication as set forth herein.

**4. Information**

You represent, warrant and covenant (with respect to Information and Projections, each as defined below, relating to the Company and its subsidiaries, to the best of your knowledge) that (i) all written information (other than Projections and information of a general economic or industry nature) (collectively, the "Information") that has been or will be made available to the Arranger or the Commitment Party by or on behalf of you in connection with the Transactions does not and will not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements contained therein not misleading in light of the circumstances under which such statements are made and (ii) the projections and other forward looking information (the "Projections") that have been or will be made available to the Arranger or the Commitment Party by or on behalf of you have been and will be prepared in good faith based upon assumptions that are believed by the preparer thereof to be reasonable when made, it being understood that any such financial projections are subject to significant uncertainties and contingencies, many of which are beyond your control, that no assurance can be given that any particular financial projections will be realized, that actual results may differ and that such differences may be material. You agree that if at any time prior to the later of (i) the Closing Date and (ii) the Syndication Date, any of the representations in the preceding sentence would be incorrect if made at such time, then you will promptly supplement, or cause to be supplemented, the Information and Projections so that such representations (to the best of your knowledge with respect to the Company and its subsidiaries) will be correct in light of the circumstances in which statements are made. You understand that in providing our services pursuant to this Commitment Letter we may use and rely on the Information and Projections without independent verification thereof.

**5. Indemnification**

To induce us to enter into this Commitment Letter and the Fee Letter and to proceed with the documentation of the Incremental Term Facility, you hereby agree to indemnify upon demand and hold harmless the Administrative Agent, the Collateral Agent, the Arranger and the Commitment Party and their respective affiliates and each partner, trustee, shareholder, director, officer, employee, advisor,

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**Jazz Pharmaceuticals Public Limited Company**

representative, agent, attorney and controlling person thereof (each of the above, an “Indemnified Person”) from and against any and all actions, suits, proceedings (including any investigations or inquiries), claims, losses, damages, liabilities or expenses (including reasonable and documented legal expenses of one firm of counsel to all Indemnified Persons, one local counsel in each appropriate jurisdiction, one special counsel and, in the case of an actual or perceived conflict of interest where the Indemnified Person affected by such conflict informs you of such conflict and thereafter retains its own counsel, of another firm of counsel for each such affected Indemnified Person), joint or several, of any kind or nature whatsoever that may be brought or threatened by the Company, the Borrower, the Guarantors, any of their respective affiliates or any other person or entity and that may be incurred by or asserted against or involve any Indemnified Person (whether or not any Indemnified Person is a party to such action, suit, proceeding or claim) as a result of or arising out of or in any way related to or resulting from the Acquisition, this Commitment Letter, the Fee Letter, the Incremental Term Facility, the Transactions or any related transaction contemplated hereby or thereby or any use or intended use of the proceeds of the Incremental Term Facility; provided that you will not have to indemnify an Indemnified Person against any claim, loss, damage, liability or expense (i) to the extent the same resulted from the gross negligence, bad faith or willful misconduct of, or a material breach of this Commitment Letter or the Fee Letter by, such Indemnified Person or any of their affiliates or its or their partners, trustees, shareholders, directors, officers, employees, advisors, representatives, agents, attorneys or controlling person (to the extent determined by a court of competent jurisdiction in a final and non-appealable judgment) or (ii) incurred in connection with any dispute among Indemnified Parties (other than a dispute against the Administrative Agent, the Collateral Agent or the Arranger in their capacities as such) other than as a result of any act or omission by the Buyer or its affiliates. Notwithstanding any other provision of this Commitment Letter, no Indemnified Person will be responsible or liable to you or any other person or entity for damages arising from the use by others of any information or other materials obtained through internet, electronic, telecommunications or other information transmission systems, except to the extent such damages have resulted from the bad faith, gross negligence or willful misconduct of, or a material breach of this Commitment Letter or the Fee Letter by, such Indemnified Person or any of its affiliates or its or their partners, trustees, shareholders, directors, officers, employees, advisors, representatives, agent, attorneys or controlling persons (in each case, to the extent determined by a court of competent jurisdiction in a final and non-appealable judgment). You shall not be liable for any settlement of any proceeding effected without your consent (which consent shall not be unreasonably withheld, conditioned or delayed), but if settled with your written consent or if there is a final and non-appealable judgment by a court of competent jurisdiction for the plaintiff in any such proceeding for which an Indemnified Person is entitled to be indemnified hereunder, you agree to indemnify and hold harmless each Indemnified Person from and against any and all losses, claims, damages, liabilities and expenses by reason of such settlement or judgment in accordance with the other provisions of this paragraph.

Your indemnity and reimbursement obligations under this Section 5 will be in addition to any liability which you may otherwise have and will be binding upon and inure to the benefit of the successors, assigns, heirs and personal representatives of you and the Indemnified Persons.

Neither (x) we nor any other Indemnified Person, nor (y) you will be responsible or liable to you or any other person or entity for any indirect, special, punitive or consequential damages (in the case of clause (y), other than in respect of any such damages required to be indemnified pursuant to this Section 5), which may be alleged as a result of the Acquisition, this Commitment Letter, the Fee Letter, the Incremental Term Facility, the Transactions or any related transaction contemplated hereby or thereby or any use or intended use of the proceeds of the Incremental Term Facility.

6. Assignments

This Commitment Letter may not be assigned by you without the prior written consent of Barclays (and any purported assignment without such consent will be null and void), is intended to be solely for the benefit of the parties hereto and is not intended to confer any benefits upon, or create any rights in favor of, any person (including your equity holders, employees or creditors) other than the parties hereto (and any Indemnified Person). Barclays may assign its commitments and agreements hereunder, in whole or in part, to any of its (i) affiliates or additional arrangers or (ii) any Lender, provided that no such assignment shall relieve Barclays of its obligations to fund such assigned portion of its commitment hereunder on the Closing Date. This Commitment Letter may not be amended or any term or provision hereof waived or modified except by an instrument in writing signed by each of the parties hereto.

7. USA PATRIOT Act Notification

Barclays notifies you, the Company, the Borrower and the Guarantors that, pursuant to the requirements of the USA PATRIOT Act (Title III of Pub. L. 107-56 (signed into law October 26, 2001)) (as amended, supplemented or modified from time to time, the "Patriot Act") it and each Lender may be required to obtain, verify and record information that identifies you, the Company, the Borrower and the Guarantors, including the name and address of each such Person and other information that will allow Barclays and each Lender to identify you, the Company, the Borrower and the Guarantors in accordance with the Patriot Act and other applicable "know your customer" and anti-money laundering rules and regulations. This notice is given in accordance with the requirements of the Patriot Act and is effective for Barclays and each Lender.

8. Sharing Information; Affiliate Activities; Absence of Fiduciary Relationship

Please note that this Commitment Letter and the Fee Letter are delivered to you on the understanding that none of this Commitment Letter or the Fee Letter or the information contained herein and therein shall be disclosed, directly or indirectly, to any other person or entity (including other lenders, underwriters, placement agents, advisors or any similar persons) without our prior written consent except pursuant to the order of any court or administrative agency or in any pending legal or administrative proceeding, or otherwise as required by applicable law or regulation or as requested by a governmental authority (in which case you agree to inform us promptly thereof to the extent lawfully permitted to do so); provided that we hereby consent to your disclosure of (i) this Commitment Letter, the Fee Letter and the information contained herein or therein to your officers, directors, employees, attorneys, accountants, agents and advisors on a confidential basis, (ii) this Commitment Letter and the information contained herein (but not the Fee Letter or the information contained therein, except to the extent redacted in a manner satisfactory to the Arranger) to the Company and its investors, officers, directors, employees, affiliates, attorneys, accountants, agents and advisors on a confidential, need to know basis, (iii) the term sheet attached as Exhibit A and the existence of this Commitment Letter (and the information contained herein) to any rating agency in connection with the Transactions, (iv) the aggregate fee amounts contained in this Commitment Letter and the Fee Letter as part of the financial statements and Projections, pro forma information or a generic disclosure of aggregate sources and uses related to fee amounts related to the Transactions to the extent customary or required in offering and marketing materials for the Incremental Term Facility or in any public filing relating to the Transactions, and (v) this Commitment Letter and the information contained herein (but not the Fee Letter and the information contained therein) in any syndication of the Incremental Term Facility and to the extent required to be filed with the U.S. Securities and Exchange Commission or other public filings in connection with the Transactions.

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**Jazz Pharmaceuticals Public Limited Company**

You acknowledge that Barclays and its affiliates are full service securities firms and as such may from time to time effect transactions, for their own account or the account of customers, and may hold positions in securities or indebtedness, or options thereon, of the Buyer, the Company and other companies that may be the subject of the Transactions. Barclays and its affiliates will have economic interests that are different from or conflict with those of the Buyer regarding the transactions contemplated hereby, and you acknowledge and agree that Barclays has no obligation to disclose such interests to you. You further acknowledge and agree that nothing in this Commitment Letter, the Fee Letter or the nature of our services or in any prior relationship will be deemed to create an advisory, fiduciary or agency relationship between us, on the one hand, and you, your equity holders or your affiliates, on the other hand, and you waive, to the fullest extent permitted by law, any claims you may have against Barclays for breach of fiduciary duty or alleged breach of fiduciary duty and agree that Barclays will have no liability (whether direct or indirect) to you in respect of such a fiduciary duty claim or to any person asserting a fiduciary duty claim on your behalf, including your equity holders, employees or creditors. You acknowledge that the Transactions (including the exercise of rights and remedies hereunder and under the Fee Letter) are arms' length commercial transactions and that we are acting as principal and in our own best interests. You are relying on your own experts and advisors to determine whether the Transactions are in your best interests and are capable of evaluating and understanding, and you understand and accept, the terms, risks and conditions of the transactions contemplated hereby. In addition, you acknowledge that we may employ the services of our affiliates in providing certain services hereunder and may exchange with such affiliates information concerning you, the Company and other companies that may be the subject of the Transactions and such affiliates will be entitled to the benefits afforded to us hereunder. In addition, please note that Barclays Capital Inc. has been retained by you as financial advisor (in such capacity, the "Financial Advisor") to you in connection with the Acquisition. You agree to such retention, and further agree not to assert any claim you might allege based on any actual or potential conflicts of interest that might be asserted to arise or result from, on the one hand, the engagement of the Financial Advisor, and on the other hand, our and our affiliates' relationships with you as described and referred to herein.

Consistent with our policies to hold in confidence the affairs of our customers, we will not use or disclose confidential information obtained from you by virtue of the Transactions in connection with our performance of services for any of our other customers (other than as permitted to be disclosed under this Section 8). Furthermore, you acknowledge that neither we nor any of our affiliates have an obligation to use in connection with the Transactions, or to furnish to you, confidential information obtained or that may be obtained by us from any other person.

Please note that Barclays and its affiliates do not provide tax, accounting or legal advice.

9. Waiver of Jury Trial; Governing Law; Submission to Jurisdiction; Surviving Provisions

ANY RIGHT TO TRIAL BY JURY WITH RESPECT TO ANY ACTION, SUIT, PROCEEDING OR CLAIM ARISING IN CONNECTION WITH OR AS A RESULT OF ANY MATTER REFERRED TO IN THIS COMMITMENT LETTER OR THE FEE LETTER IS HEREBY IRREVOCABLY WAIVED BY THE PARTIES HERETO. THIS COMMITMENT LETTER WILL BE GOVERNED BY AND CONSTRUED IN ACCORDANCE WITH THE LAWS OF THE STATE OF NEW YORK; *provided, however*, that each of (a) the interpretation of the definition of Company Material Adverse Effect and whether there shall have occurred a Company Material Adverse Effect and (b) whether the Initial Acquisition has been consummated in accordance with the terms of the Acquisition Agreement shall be governed and construed in accordance with the laws of the State of Delaware applicable to agreements made and to be performed entirely within such State, without regard to the conflict of laws principles of such State to the extent that the laws of another jurisdiction would be required thereby. Each of the parties hereto hereby irrevocably (i) submits, for itself and its property, to the exclusive jurisdiction of (a) the Supreme Court of the State of New York, New York County, located in the Borough of Manhattan and (b) the United States District Court for the Southern District of New York and any appellate court



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**Jazz Pharmaceuticals Public Limited Company**

from any such court, in any action, suit, proceeding or claim arising out of or relating to this Commitment Letter, the Fee Letter or the Transactions or the performance of services contemplated hereunder or under the Fee Letter, or for recognition or enforcement of any judgment, and agrees that all claims in respect of any such action, suit, proceeding or claim may be heard and determined in such New York State court or such Federal court, (ii) waives, to the fullest extent permitted by law, any objection that it may now or hereafter have to the laying of venue of any action, suit, proceeding or claim arising out of or relating to this Commitment Letter, the Fee Letter, the Transactions or the performance of services contemplated hereunder or under the Fee Letter in any such New York State or Federal court and (iii) waives, to the fullest extent permitted by law, the defense of an inconvenient forum to the maintenance of any such action, suit, proceeding or claim in any such court. Each of the parties hereto agrees to commence any such action, suit, proceeding or claim either in the United States District Court for the Southern District of New York or in the Supreme Court of the State of New York, New York County located in the Borough of Manhattan. The Buyer hereby irrevocably appoints Jazz U.S. as its agent for service of process for purpose of the submission to jurisdiction set forth above.

This Commitment Letter is issued for your benefit only and no other person or entity (other than the Indemnified Persons) may rely hereon.

The provisions of Sections 3, 5, 6, 8 and this Section 9 of this Commitment Letter will survive any termination or completion of the arrangements contemplated by this Commitment Letter or the Fee Letter, including without limitation whether or not the Incremental Amendment is executed and delivered and whether or not the Incremental Term Facility are made available or any loans under the Incremental Term Facility are disbursed, provided that (i) the provisions of Section 3 shall survive until the earlier of termination of this Commitment Letter (provided that there has been no initial funding of the Incremental Term Facility) or the Syndication Date and (ii) the provisions of Section 5 shall be superseded by the definitive documentation for the Incremental Term Facility to the extent covered thereby.

10. Termination; Acceptance

Our commitments hereunder and our agreements to provide the services described herein will terminate upon the first to occur of (i) the consummation of the Initial Acquisition (as defined in Exhibit B) without borrowings under the Incremental Term Facility, (ii) the abandonment or termination of the definitive documentation for the Acquisition, including a valid termination of the Acquisition Agreement after execution thereof and prior to the consummation of the Initial Acquisition, (iii) April 19, 2014 (or, to the extent the Termination Date (as defined in the Acquisition Agreement on the date hereof) has been extended in accordance with Section 7.1(b) of the Acquisition Agreement, June 19, 2014) unless the closing of the Incremental Term Facility has been consummated on or before such date on the terms and subject to the conditions set forth herein, and (iv) the termination of this Commitment Letter and the commitments hereunder by you.

This Commitment Letter may be executed in any number of counterparts, each of which when executed will be an original and all of which, when taken together, will constitute one agreement. Delivery of an executed counterpart of a signature page of this Commitment Letter by facsimile or other electronic transmission will be as effective as delivery of a manually executed counterpart hereof.

Please confirm that the foregoing is in accordance with your understanding by signing and returning to Barclays the enclosed copy of this Commitment Letter, together, if not previously executed and delivered, with the Fee Letter on or before the close of business on December 20, 2013, whereupon this Commitment Letter and the Fee Letter will become binding agreements between us. If not signed and returned as described in the preceding sentence by such date, this offer will terminate on such date.

*[The remainder of this page is intentionally left blank.]*

We look forward to working with you on this assignment.

Very truly yours,

**BARCLAYS BANK PLC**

By: /s/ Ian Palmer

Name: Ian Palmer

Title: Managing Director

Commitment Letter

ACCEPTED AND AGREED TO AS OF THE DATE FIRST WRITTEN ABOVE:

JAZZ PHARMACEUTICALS PUBLIC LIMITED COMPANY

By: /s/ Fintan Keegan

Name: Fintan Keegan

Title: Executive Vice President

Commitment Letter

**Summary of Terms and Conditions of the Incremental Term Facility**

*This Summary of Terms and Conditions outlines certain terms and conditions of the Incremental Term Facility.*

- Borrower:** Jazz Pharmaceuticals, Inc., a Delaware corporation (“Jazz U.S.” or the “Borrower”).
- Guarantors:** The same as under the Existing Credit Agreement, it being understood that on the Closing Date, the Borrower shall cause the Company and its subsidiaries to become Guarantors to the extent required by the terms of the Existing Credit Agreement (the “New Guarantors”).
- Sole Bookrunner and Sole Lead Arranger:** Barclays Bank PLC will act as sole bookrunner and sole lead arranger (in such capacities, the “Arranger”) for the Incremental Term Facility and will perform the duties customarily associated with such roles.
- Administrative Agent:** Barclays Bank PLC (in such capacity, the “Administrative Agent”).
- Collateral Agent:** Barclays Bank PLC (in such capacity, the “Collateral Agent”).
- Lenders:** Barclays Bank PLC and/or other banks, financial institutions and institutional lenders selected by the Arranger (each, a “Lender” and, collectively, the “Lenders”).
- Purpose/Use of Proceeds:** The proceeds of the Incremental Term Facility will be used to fund, together with cash on hand, the Acquisition and the Transaction Costs.
- Amount of Incremental Facility:** A senior secured incremental term loan facility in an aggregate principal amount of \$500 million (the “Incremental Term Facility”).
- Definitive Documentation:** The Incremental Term Facility shall be documented as a new tranche of term loans under the Existing Credit Agreement, subject to the same terms and provisions contained therein, with such modifications as set forth herein and in the Incremental Amendment with respect thereto.
- Incremental Facilities:** Same as the Existing Credit Agreement, it being understood that the Incremental Term Facility shall reduce, or otherwise be deemed to be a use of, the borrowing capacity under the incremental facilities of the Existing Credit Agreement. The Incremental Term Facility will have the benefit of the 50 basis points “most favored nation” provision applicable to the existing Term Loans under the Existing Credit Agreement.

<b>Borrower:</b>	Jazz Pharmaceuticals, Inc., a Delaware corporation (“ <u>Jazz U.S.</u> ” or the “ <u>Borrower</u> ”).
<b>Refinancing Facilities:</b>	Same as the Existing Credit Agreement.
<b>Availability:</b>	One drawing may be made under the Incremental Term Facility on the Closing Date.
<b>Closing Date:</b>	The date on which the borrowings under the Incremental Term Facility are made and the Initial Acquisition is consummated in accordance with the Commitment Letter (the “ <u>Closing Date</u> ”).
<b>Maturities:</b>	June 12, 2018 (the same as the Tranche 1 Term Loans under the Existing Credit Agreement) (the “ <u>Maturity Date</u> ”).
<b>Amortization:</b>	The outstanding principal amount of the Incremental Term Facility will be payable in equal quarterly amounts of 1.0% <i>per annum</i> prior to the Maturity Date, commencing with the first Principal Amortization Payment Date following the Closing Date, with the remaining balance, together with all other amounts owed with respect thereto, payable on the Maturity Date.
<b>Interest Rate:</b>	All amounts outstanding under the Incremental Term Facility will bear interest, at the Borrower’s option, at a rate <i>per annum</i> equal to: <ul style="list-style-type: none"> <li>(a) the Base Rate plus 1.75% <i>per annum</i>; or</li> <li>(b) the Adjusted Eurodollar Rate <u>plus</u> 2.75% <i>per annum</i>;</li> </ul> <u>provided</u> , that at no time will the Base Rate be deemed to be less than 1.75% <i>per annum</i> or the reserve adjusted Eurodollar Rate be deemed to be less than 0.75% <i>per annum</i> .
<b>Default Interest:</b>	Same as the Existing Credit Agreement.
<b>Funding Protection and Taxes:</b>	Same as the Existing Credit Agreement.
<b>Closing Fees:</b>	Closing fees to each Lender party to the Incremental Amendment on the Closing Date, as fee compensation for the funding of such Lender’s loans under the Incremental Term Facility, in an amount equal to 0.50% of the stated principal amount of such Lender’s loans under the Incremental Term Facility, payable to such Lender on the Closing Date.
<b>Voluntary Prepayments:</b>	Same as the Existing Credit Facility; provided that upon the occurrence of any Repricing Transaction (as defined in the Existing Credit Agreement) prior to the date that is six months after the Closing Date, the Borrower shall be required to pay a fee in the amount equal to 1.00% of the principal amount of the loans under the Incremental Term Facility subject to such Repricing Transaction.

<b>Borrower:</b>	Jazz Pharmaceuticals, Inc., a Delaware corporation (“ <u>Jazz U.S.</u> ” or the “ <u>Borrower</u> ”).
<b>Mandatory Prepayments:</b>	Same as the Existing Credit Facility. The loans under the Incremental Term Facility will share ratably in all mandatory prepayments with the existing Term Loans under the Existing Credit Agreement.
<b>Collateral:</b>	Same as the Existing Credit Agreement.
<b>Representations and Warranties:</b>	Same as the Existing Credit Agreement.
<b>Affirmative Covenants:</b>	Same as the Existing Credit Agreement.
<b>Negative Covenants:</b>	Same as the Existing Credit Agreement.
<b>Financial Covenants:</b>	Same as the Existing Credit Facility.
<b>Events of Default:</b>	Same as the Existing Credit Agreement.
<b>Margin Regulation Savings Clauses:</b>	The Incremental Amendment will provide that, for so long as Company Stock constitutes “margin stock” within the meaning of Regulation U, the restrictions on negative pledges, liens and dispositions, the mandatory prepayment provisions and the related cross default provisions set forth in the Existing Credit Agreement shall not apply with respect to Company Stock to the extent the value (within the meaning of Regulation U) of Company Stock, together with the value of all other margin stock held by the Borrower and its subsidiaries, exceeds 25% of the total value of all assets subject to such covenants and agreements.
<b>Conditions Precedent to Borrowing:</b>	The several obligation of each Lender to make, or cause an affiliate to make, loans under the Incremental Term Facility on the Closing Date will be subject only to (i) prior written notice of borrowing, (ii) satisfaction of the conditions set forth in Section 4.02(b) and (c) of the Existing Credit Agreement, (iii) after giving effect to the making of loans under the Incremental Term Facility, Parent shall be in compliance with the covenant set forth in Section 7.10 of the Existing Credit Agreement on a pro forma basis in accordance with Section 1.03(c) of the Existing Credit Agreement and (iv) satisfaction of the conditions set forth in Exhibit B to the Commitment Letter.
<b>Assignments and Participations:</b>	Same as the Existing Credit Agreement.

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**Borrower:** Jazz Pharmaceuticals, Inc., a Delaware corporation (“Jazz U.S.” or the “Borrower”).

**Amendments and Required Lenders:** Same as the Existing Credit Agreement.

**Indemnity and Expenses:** Same as the Existing Credit Agreement.

**Governing Law and Jurisdiction:** Same as the Existing Credit Agreement.

**Counsel to the Arranger and the Administrative Agent:** Cahill Gordon & Reindel LLP.

## Exhibit B

### Summary of Conditions Precedent to the Incremental Term Facility

1. Current Transactions: The terms of the Acquisition Agreement (including all exhibits, schedules, annexes and other attachments thereto) and all related documents shall be reasonably satisfactory to the Arranger (it being agreed that the draft Acquisition Agreement and related documents and schedules provided to the Arranger on December 19, 2013 are reasonably satisfactory to the Arranger). The acquisition, pursuant to the Tender Offer, of more than 50% of the outstanding Company Stock (the "Initial Acquisition") shall have been consummated or will be consummated substantially concurrently with the initial funding under the Incremental Term Facility in accordance with the terms of the Acquisition Agreement; provided that no amendment, modification or waiver of any term of the Acquisition Agreement or any condition to the Buyer or its subsidiary's obligation to consummate the Acquisition thereunder (other than any such amendment, modification or waiver that is not materially adverse to any interest of the Lenders and other than a Permitted Minimum Condition Modification (as defined in the Acquisition Agreement)) shall be made or granted, as the case may be, without the prior written consent of the Commitment Party (it being understood that any reduction in the price that is less than or equal to 10% of the total consideration set forth in the Acquisition Agreement as of the date hereof will not be deemed to be materially adverse to the interests of the Lenders and will not require the prior written consent of the Commitment Party).
2. Financial Statements. The Arranger shall have received (i) unqualified audited financial statements of the Company for its three most recently completed fiscal years ended 90 days before the Closing Date (it being understood that the Arranger has received such financial statements for each of the fiscal years ended December 31, 2010, 2011 and 2012) and (ii) unaudited financial statements for any quarterly interim period or periods of the Company ending more than 45 days prior to the Closing Date (it being understood that the quarter ending December 31, 2013 is not an interim period), together with unaudited financial statements for the corresponding period of the prior year (it being understood that the Arranger has received such financial statements for the quarterly periods ended March 31, 2013, June 30, 2013 and September 30, 2013); provided that the filing of the required financial statements with the U.S. Securities and Exchange Commission will satisfy the foregoing requirements. All such financial statements shall have been prepared in accordance with U.S. GAAP (except, in the case of the interim financials, for year-end audit adjustments and absence of footnotes).
3. Marketing Period. The Arranger shall have been afforded a period of no less than 15 consecutive calendar days following the date that the Marketing Materials have been provided to syndicate the Incremental Term Facility prior to the Closing Date; *provided* that such 15 consecutive calendar day period shall not commence earlier than January 6, 2014.
4. No Material Adverse Effect. As of the scheduled expiration date of the Offer (as defined in the Acquisition Agreement), since June 30, 2013 there shall not have occurred a Company Material Adverse Effect (as defined in the Acquisition Agreement on the date hereof), and there has not been, and there does not exist, any Effect (as defined in the Acquisition Agreement on the date hereof) that, individually or in the aggregate with other Effects, would reasonably be expected to have a Company Materially Adverse Effect.
5. Fees and Expenses. All fees and expenses (in the case of expenses, to the extent invoiced at least three business days prior to the Closing Date) required to be paid on the Closing Date pursuant to the Commitment Letter and the Fee Letter shall, upon the initial borrowing under the Incremental Term Facility, have been paid (which amounts may be offset against the proceeds of the Incremental Term Facility).



6. Definitive Documents; Customary Closing Conditions. The Borrower and the Guarantors shall have executed definitive loan documents relating to the Incremental Term Facility, including without limitation, the Incremental Amendment, guarantees, security agreements, pledge agreements, real property security agreements, or in each case, to the extent applicable, amendments or joinders thereto, and other related definitive documents (collectively, the "Loan Documents"), which shall be consistent with the terms set forth in this Commitment Letter and subject to the Conditions Limitation Provision and otherwise reasonably satisfactory to the Commitment Party. Subject to the Conditions Limitation Provision, the Arranger shall be reasonably satisfied that the Borrower has complied with all other customary closing conditions, including without limitation: (i) the delivery of customary legal opinions, corporate records and documents from public officials, officer's certificates and evidence of authority; and (ii) delivery of a solvency certificate (certifying that, after giving effect to the Transactions, the Parent and its subsidiaries on a consolidated basis are solvent) in substantially the form of Exhibit K to the Existing Credit Agreement.
7. Patriot Act. The Administrative Agent and the Arranger shall have received at least five days prior to the Closing Date, all documentation and other information about the Borrower and the Guarantors (including the New Guarantors) as required by regulatory authorities under applicable "know your customer" and anti-money laundering rules and regulations, including without limitation the Patriot Act, to the extent reasonably requested by any Lender to the Administrative Agent and conveyed by the Administrative Agent to the Borrower in writing at least 10 days prior to the Closing Date.

**FORM OF  
TRANSITION, AMENDMENT AND RELEASE AGREEMENT**

*between*

**Gentium GmbH**, General-Guisan-Strasse 6, 6303 Zug, Switzerland

(hereinafter the “Company”)

*and*

[—]

(hereinafter the “Employee”)

WHEREAS, the Employee is currently employed by the Company pursuant to an employment agreement with the Company (the employment agreement as amended referred to as “Employment Agreement”);

WHEREAS, Gentium S.p.A. intends to enter into a Tender Offer Agreement (the “TO Agreement”) by and among Jazz Pharmaceuticals plc, a public limited company formed under the laws of Ireland (the “Parent”), Jazz Pharmaceuticals Italy S.r.l., a corporation and a wholly owned subsidiary of Parent (the “Purchaser”), and Gentium S.p.A. on or around December 19, 2013 (the “TO Agreement Date”). Following the TO Agreement Date the Purchaser shall commence a tender offer (the “Offer”) to acquire all of the outstanding shares of Gentium S.p.A.’s common stock and Gentium S.p.A.’s ADSs (together, the “Shares”). On the terms set forth in the TO Agreement, Purchaser shall accept for payment, and pay for, all Shares validly tendered and not withdrawn pursuant to the Offer, promptly after the applicable expiration date of the Offer (the “Closing Date”);

WHEREAS, the Employee, as one of the principal shareholders of Gentium S.p.A has a vital interest in the consummation of the Offer;

WHEREAS, the Employee has indicated that he would like to resign as an as an employee of the Company if the Offer is consummated;

NOW, therefore, in consideration of the mutual covenants and agreements set forth hereinafter, the parties agree as follows:

1. The Employee herewith resigns on the Closing Date (the "Resignation Date") and the parties herewith agree that the Employment Agreement and the employment relationship existing between them shall terminate as of the Resignation Date.
2. Until the Resignation Date, the Employee will continue to receive compensation as provided for in his Employment Agreement.
3. The parties agree to amend the Employee's Employment Agreement such that he will only be eligible to receive the entitlements specified under Section 11.3 if he successfully completes his Consultancy Agreement as determined by the Company in its discretion.
4. As such, and subject to paragraph 3, provided that the Employee signs a waiver and release substantially in the form attached hereto as Exhibit A no earlier than one (1) full month after the Resignation Date and no later than two (2) months after the Resignation Date and successfully completes the Consultancy Agreement, and he signs a subsequent waiver and release no earlier than one (1) full month after the end date of the Consultancy Agreement and no later than two (2) months after the end date of the Consultancy Agreement also in a form substantially in the form attached hereto as Exhibit A, then the Company will provide to the Employee all entitlements pursuant to Section 11.3 of his Employment Agreement in full settlement of all claims and rights of action (whether under statute, contract, common law or otherwise) that the Employee

might have in connection with his employment and its termination or any other matter against the Company or any undertaking affiliated to the Company or its or their officers, employees or shareholders, in any jurisdiction in the world.

5. The Employee shall, on the Resignation Date, hand over to the Company the company car and all such other documents and items belonging to the Company that the Company may request. Any retention right is excluded.
6. During the term of the Employment Agreement and at any time thereafter, the Employee shall keep strictly confidential and neither use for his own purposes or that of others nor make known to any third person any knowledge or data obtained by him during his employment with the Company and/or Gentium S.p.A., including but not limited to: information related to the business of the Company and/or any company affiliated with the Company, information regarding inventions, research or development activities and plans, compounds and biological materials, products, specifications, manufacturing, production and testing processes, systems, methods and techniques, costs of production, sales, marketing and promotional information and plans, lists of names or classes of suppliers or customers, personnel, business plans and strategy, financial statements and information and the existence and content of any business discussions, negotiations or agreements between the Company and/or any company affiliated with the Company and any third party.
7. The Employee herewith transfers to the Company all intellectual property rights including but not limited to inventions, manufacturing, production, industrial and testing processes, systems, methods, procedures and techniques, improvements, trade secrets, software programs, software and systems documentation, works of authorship and other copyrightable works, and related know-how which result from work performed by Employee, alone or with others, whether or not patentable, copyrightable, or qualified for other protection as proprietary information ensuing from any work performed by the Employee during the term of his employment with the Company and/or Gentium S.p.A. and any concepts, in written or by any other means machine-readable form

("Intellectual Property Rights"). The Employee may not, without the Company's written consent, disclose, multiply, use, manufacture, bring on the market or sell, lease, deliver or otherwise trade, offer, or register the results of his work. The Employee agrees, furthermore, to sign any documents and to take any measures that the Company may request to effect the transfer of the Intellectual Property Rights and full and exclusive title of the Company in the Intellectual Property Rights provided. The Company is entitled to transfer the Intellectual Property Rights in full or in part to any third party. The Company and such third parties are not obliged to mention the Employee as the author if they publish any inventions, computer programs or other works. They are free to make any modifications, translations and/or other adaptations and/or can refrain from making any publications.

8. The Company herewith informs the Employee that his accident insurance will end 30 days after the Resignation Date. If he has not commenced new employment that provides for insurance coverage of the occupational as well as non-occupational accident, the Employee has to notify his health insurer about the fact that the employment relationship will end on the Resignation Date to secure sufficient insurance coverage or has to take out a so-called "Abrediversicherung" with the current accident insurance company for up to 180 days. If the Employee is interested in such insurance coverage he shall contact the Company's Human Resources Department.
9. The Employee will not in any way disparage or criticize the Company or any company affiliated to the Company, their respective directors, officers or employees or any of the Company's products or those of any other company affiliated to the Company. Similarly, the Company's director, officers and executives shall not in any way disparage or criticize the Employee.
10. Both parties hereby release and forever discharge the other party and their related entities and their respective directors, officers, stockholders, employees, agents or attorneys of and from any and all claims of any nature whatsoever.

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This release does not include the obligations of the parties arising out of this Agreement or the TO Agreement.

11. The terms of this Agreement constitute the entire agreement and understanding between the parties hereto and it supersedes and replaces all prior negotiations, agreements, arrangements, or understandings (whether implied or expressed, oral or in writing) concerning the subject matter hereof, all of which are hereby treated as terminated by mutual consent.
12. Modifications of and amendments to this Agreement, including this section 12, shall exclusively be made in writing.

13. The present Agreement shall be governed by Swiss law, in particular art. 319 ss. of the Swiss Code of Obligations.

Place and date

Place and date

The Company

The Employee

**Gentium GmbH**

[—]

**WAIVER AND RELEASE**

The undersigned, [—], herewith waives any and all claims and rights of action (whether under statute, contract, common law or otherwise) that he has or might have against Gentium S.p.A. and Gentium GmbH or any undertaking affiliated to Gentium S.p.A., Gentium GmbH, or its parent, any subsidiaries or affiliates or their officers, employees or shareholders, in any jurisdiction in the world, in particular those arising out of or in connection with his employment or appointment with Gentium S.p.A. and Gentium GmbH and its termination or any other matter.

This release does not include the obligations of the parties arising out of the Termination, Amendment and Transition Agreement or the Tender Offer Agreement.

\_\_\_\_\_  
Date

\_\_\_\_\_  
[—]



**FORM OF  
CONSULTANCY AGREEMENT**

*between*

**Gentium S.p.A., Piazza XX Settembre 2, 22079 Villa Guardia (Como), Italy**

(hereinafter "Gentium")

*and*

[—]

(hereinafter the "Consultant")

1. Contingent upon the successful close of the tender offer by Jazz Pharmaceuticals Italy S.r.l. to acquire all of the outstanding shares of Gentium's common stock and Gentium's ADSs, the Consultant agrees to render the following services to Gentium beginning on the date on which he resigns as an employee of the Company ("Commencement Date"): provide his knowledge and expert opinion in the form of meetings, advice and the like, as will be agreed to by the parties in advance, as they relate to the products, technology, strategy and long-term success of the business of Gentium and the therapeutic areas in which it currently operates or may desire to operate ("Services").
2. The Consultant shall not, by virtue of his activity as Consultant to Gentium, be deemed an employee, a partner or a shareholder of Gentium and shall, to the extent that no specific written power of attorney has been granted for an individual case, not be entitled to represent Gentium in a legally binding manner.

3. It is anticipated that Consultant shall provide these Services for six (6) months. During the first month, Consultant shall provide Services as requested by Gentium for up to two (2) days per week. After the first month, and for the remaining five (5) months, Consultant shall provide Services as requested by Gentium for up to one (1) day per week.
4. The Consultant shall be compensated for his performance hereunder in a monthly retainer of EUR 12,000 for the first month of Services, followed by a monthly retainer of EUR 6,000 for the remaining five (5) months. Such compensation shall cover any work performed hereunder as well as any acquisition of rights under Article 7.
5. Gentium shall not be liable for any taxes other than VAT and duties on such compensation. In particular, the Consultant undertakes to account for and pay himself to the pertinent social security authorities all contributions he owes due to services performed under this Agreement in a timely manner and shall indemnify Gentium in case any claims for payment of social security contributions, taxes and duties are made by any Swiss or foreign authority. He shall, upon Gentium's request, submit proof of his recognition as a self-employed by the pertinent social security authority and the payment of the pertinent social security contributions. In case the Consultant fails to submit such proof within ten (10) days upon Gentium's request, Gentium shall be entitled to withhold the entire social security contributions due from the fee payable to the Consultant.
6. During the term of this Agreement and at any time thereafter the Consultant shall keep strictly confidential and neither use for his own purposes or that of others nor make known to any third person any information, knowledge or data obtained by him during the term of this Agreement with respect to Gentium or any company affiliated to Gentium including but not limited to information related to the business of Gentium or any company affiliated with Gentium, information regarding inventions, research or development activities and plans, compounds and biological materials, products, specifications, manufacturing, production and testing processes, systems, methods and techniques, costs of production, sales, marketing and promotional information and plans, lists of

names or classes of suppliers or customers, personnel, business plans and strategy, financial statements and information and the existence and content of any business discussions, negotiations or agreements between Gentium or any company affiliated with Gentium and any third party, trade secrets, or know-how or data which at such time of disclosure is not publicly known and in the public domain, regardless of whether such information is confidential.

7. The Consultant agrees that any work products originating from the performance of the duties and tasks hereunder, including but not limited to inventions, manufacturing, production, industrial and testing processes, systems, methods, procedures and techniques, improvements, trade secrets, software programs, software and systems documentation, works of authorship and other copyrightable works, and related know-how which result from work performed by Consultant, alone or with others, whether or not patentable, copyrightable, or qualified for other protection as proprietary information, and any concepts, in written or by any other means machine-readable form shall inure to the full benefit of Gentium, and the Consultant, therefore, assigns any rights pertaining to such work products to the extent legally permissible to Gentium, and further agrees that Gentium subsequently may assign any such rights. With respect to any rights which as a matter of law remain the intellectual property of the Consultant, the Consultant herewith grants Gentium an exclusive and transferable, royalty free license, unlimited as to time and territory, for all uses, including the right to assign or sublicense such right to any third party. The license includes, among others, the right to publish the work products or parts of it on an electronic system, in a book or other publication; and to translate or otherwise modify the work product.
8. This Agreement shall terminate six (6) months after the Commencement Date and can be terminated by either party at any time upon one (1) month's prior written notice. Upon termination, the Consultant shall immediately hand over to Gentium any documentation relating to the work products created and the performance of the tasks hereunder, and shall have no right to retain any such documentation.

9. The terms of this Agreement constitute the entire agreement and understanding between the parties hereto and it supersedes and replaces all prior negotiations, agreements, arrangements, or understandings (whether implied or expressed, oral or in writing) concerning the subject matter hereof, all of which are hereby treated as terminated by mutual consent.
10. Modifications of and amendments to this Agreement, including this section 10, shall exclusively be made in writing.
11. Should any provision herein be deemed invalid or unenforceable, such provision shall be replaced by a provision as close as legally possible to the parties' original intention. Any declaration of invalidity or unenforceability of a provision herein shall not affect the validity and enforceability of the remainder herein.
12. The present Agreement shall be governed by Swiss law. Exclusive place of jurisdiction shall be Zug, Switzerland.

Place and date

Place and date

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Gentium S.p.A.

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The Consultant

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[—]

**FORM OF  
RETENTION AND AMENDMENT AGREEMENT**

*between*

**Gentium GmbH**, General-Guisan-Strasse 6, 6303 Zug, Switzerland

(hereinafter the "Company")

*and*

\_\_\_\_\_, \_\_\_\_\_, \_\_\_\_\_  
(hereinafter the "Employee")

WHEREAS, the Employee is currently employed by the Company pursuant to an employment agreement concluded between the Employee and Gentium S.p.A on \_\_\_\_\_, as amended by \_\_\_\_\_ (the employment agreement as amended referred to as "Employment Agreement");

WHEREAS, Gentium S.p.A. intends to enter into a Tender Offer Agreement (the "TO Agreement") by and among Jazz Pharmaceuticals plc, a public limited company formed under the laws of Ireland (the "Parent"), Jazz Pharmaceuticals Italy S.r.l., a corporation and a wholly owned subsidiary of Parent (the "Purchaser"), and Gentium S.p.A. on or around December 19, 2013 (the "TO Agreement Date"). Following the TO Agreement Date the Purchaser shall commence a tender offer (the "Offer") to acquire all of the outstanding shares of Gentium S.p.A's common stock and Gentium S.p.A's ADSs (together, the "Shares"). On the terms set forth in the TO Agreement, Purchaser shall accept for payment, and pay for, all Shares validly tendered and not withdrawn pursuant to the Offer, promptly after the applicable expiration date of the Offer (the "Closing Date");

WHEREAS, the Employee, as a shareholder of the Company's parent company, is interested in the consummation of the Offer and the continued employment by the Company for a period of time after the Closing Date;

NOW, therefore, in consideration of the mutual covenants and agreements set forth hereinafter, the parties agree as follows:

1. Provided that the Employee is still employed by the Company, has worked to the satisfaction of the Company and provided further that neither party has given notice of termination months following the Closing Date ("Retention Period"), the Company shall pay to the Employee a gross amount of CHF as a retention bonus (the "Retention Bonus"). If the Company terminates the employment during the Retention Period with Cause or the Employee resigns, the Company shall not make any payment under this section 1. If the Company terminates the employment during the Retention Period without Cause (as defined within the Employment Agreement), the Company shall pay the Retention Bonus to Employee under this section 1.
2. Section 11.6 of the Employment Agreement is amended as follows:
  - 11.6 Where a Change In Control occurs (as defined below), and your employment is terminated by the Company without Cause (as defined below) or you resign for Good Reason (as defined below) within 24 months of the date of the Change In Control, you will be entitled to receive severance pay equal to (a) an amount equal to the aggregate of (i) 1.5 times your Base Salary in effect at such time of termination, (ii) 1.5 times the greater of the average of the Bonuses received in the last 3 years of employment or the targeted Bonus for the year in which you are terminated or resign and (iii) the prorated targeted Bonus for the year in which you are terminated or resign as accrued in the course of the year of termination or resignation; provided, however, that if you resign for Good Reason prior to completion of your Retention Period (as defined in the Retention and Amendment Agreement entered into by the parties hereto as of December 19, 2013), you hereby waive your rights to, and will not be entitled to, receive such severance pay.

For the avoidance of doubt, the Employee's agreement to remain employed by the Company during the Retention Period shall not constitute a waiver of his entitlement to resign for Good Reason and receive a severance payment in accordance with section 11.6 of the Employment Agreement as amended by this Agreement, after completion of the Retention Period.

3. During the term of the Employment Agreement and at any time thereafter the Employee shall keep strictly confidential and neither use for his own purposes or that of others nor make known to any third person any knowledge or data obtained by him during his employment with the Company and/or Gentium S.p.A., including but not limited to information related to the business of the Company and companies affiliated with the Company, information regarding inventions, research or development activities and plans, compounds and biological materials, products, specifications, manufacturing, production and testing processes, systems, methods and techniques, costs of production, sales, marketing and promotional information and plans, lists of names or classes of suppliers or customers, personnel, business plans and strategy, financial statements and information and the existence and content of any business discussions, negotiations or agreements between the Company and/or any company affiliated with the Company and any third party.
4. All intellectual property rights shall exclusively vest in the Company, including but not limited to inventions, manufacturing, production, industrial and testing processes, systems, methods, procedures and techniques, improvements, trade secrets, software programs, software and systems documentation, works of authorship and other copyrightable works, and related know-how which result from work performed by Employee, alone or with others, whether or not patentable, copyrightable, or qualified for other protection as proprietary information ensuing from any work performed by the Employee during the term of his employment with the Company and /or any company affiliated with the Company and any concepts, in written or by any other means machine-readable form (hereinafter the "Intellectual Property Rights"). The Employee may not, without the Company's written consent, disclose, multiply, use, manufacture, bring on the market or sell, lease, deliver or otherwise trade, offer, or register the results of his work.

Insofar as Intellectual Property Rights are not vested in the Company by operation of law or based on the first sentence of this section 4, the Employee covenants that he will transfer and, insofar as possible, hereby transfers to the Company such Intellectual Property Rights. This also applies to Intellectual Property Rights that already exist at the time this Agreement is signed. If a transfer should not be possible under the applicable law, then the Employee shall grant to the Company a worldwide, perpetual, transferable, sub-licensable, royalty-free license to use and exploit such Intellectual Property Rights in any way the Company sees fit.

The Company is entitled to transfer the Intellectual Property Rights in full or in part to any third party. The Company and such third parties are not obliged to mention the Employee as the author if they publish any inventions, computer programs or other works. They are free to make any modifications, translations and/or other adaptations and/or can refrain from making any publications.

5. The Employee's receipt of the Retention Bonus is conditioned on the successful consummation of the Offer subject to the terms of the TO Agreement. For the avoidance of doubt, if the Offer is terminated or withdrawn pursuant to the terms of the TO Agreement, this Retention and Amendment Agreement and the amendment to the Employment Agreement contained herein shall be null and void and the Employee shall not be entitled to the Retention Bonus.
6. Unless explicitly provided differently in this Agreement, the terms of the Employment Agreement shall remain in full force and effect.
7. Modifications of and amendments to this Agreement, including this section 7, shall exclusively be made in writing.



8. The present Agreement shall be governed by Swiss law, in particular art. 319 ss. of the Swiss Code of Obligations.

Place and date

Place and date

The Company

The Employee

**Gentium GmbH**

[ — ]



## CONFIDENTIALITY AND NONDISCLOSURE AGREEMENT

THIS AGREEMENT is made and entered into effective September 30, 2013 (the “Effective Date”) by and between **Gentium S.p.A.** (“Gentium”) having its principal office at Piazza XX Settembre 2, 22079—Villa Guardia (CO), Italy, and **Jazz Pharmaceuticals plc**, an Irish company having an office at Fourth Floor, Connaught House, One Burlington Road, Dublin 4, Ireland (each, a “Party” and collectively, the “Parties”).

Whereas, the Parties are interested in discussing and evaluating a possible business or technical collaborative relationship, acquisition or other extraordinary transaction pertaining to the pharmaceutical business (or part thereof) of Gentium and/or its affiliates, and each Party may disclose to the other, in connection with these discussions and evaluation (the “Purpose”), certain information that it believes to be proprietary and confidential, and neither Party is willing to proceed with these discussions unless they have an agreement restricting further disclosure or use of this information.

NOW, THEREFORE, in consideration of the mutual promises contained herein, the Parties agree as follows:

1. Definition of Confidential Information. The term “Confidential Information” as used in this Agreement shall mean any and all business information related to the plans, prospects, products, properties, finances and operations of Disclosing Party (as defined below), and includes, without limitation, technical and non-technical information, whether relating to intellectual property, such as patents, copyrights, trade secrets, proprietary information, techniques, inventions, know how, or processes, and information about one of their businesses, such as research, experimental work, development, design details and specifications, financial information, manufacturing, business forecasts, employee capabilities, sales and merchandising and marketing plans and information. “Confidential Information” also includes proprietary or confidential information of any third party that is disclosed to a Party in the course of such Party’s business and the fact and terms of any discussions between the Parties with respect to the Purpose. Any information disclosed by the disclosing party (the “Disclosing Party”) to the receiving party (the “Receiving Party”) on or after the Effective Date will be considered Confidential Information of Disclosing Party by the Receiving Party whether or not such information is designated as “Confidential” or “Proprietary”, or identified during the course of an oral disclosure as confidential, if the Receiving Party should know by virtue of the circumstances in which Receiving Party learned the information, including through visual or other inspection, that such information is not generally known to the public or to anyone who is not bound to the Disclosing Party by obligations of confidentiality. The Parties agree that this Agreement is not intended to restrict use or disclosure of any portion of such Confidential Information which:
  - a. Is now or later made known to the public through no default by the Receiving Party of its obligations under this Agreement;



- b. The Receiving Party can show was in its possession prior to the earliest disclosure by the Disclosing Party;
  - c. Is received by the Receiving Party from a third party which, to the Receiving Party's knowledge after due inquiry, was not prohibited from disclosing such information to the Receiving Party by any obligation of confidentiality to the Disclosing Party;
  - d. Is independently developed by the Receiving Party independent of, and without any reference to, Confidential Information of the Disclosing Party; or
  - e. Is disclosed by the Receiving Party after receipt of written permission from the Disclosing Party.
2. **Obligation of Confidentiality.** A Receiving Party agrees to hold in confidence and, except as otherwise permitted by this Agreement, not to publish or disclose to any third parties any of the Confidential Information of the Disclosing Party without the prior written consent of the Disclosing Party. Each Party agrees to use the same degree of care (and in any event not less than reasonable care) to safeguard the confidentiality of the Confidential Information that it uses to protect its own secret or confidential information of a similar nature. A Receiving Party agrees to limit any disclosure of the Confidential Information only to those of its employees, independent contractors and outside professional advisors who have a need to know and who are bound by confidentiality obligations, and to advise such persons of the Receiving Party's obligations under this Agreement.
3. **Use of Confidential Information.** Each Party agrees to use Confidential Information received from the other Party only:
- a. For the Purpose, and
  - b. To pursue a technical or business collaborative relationship after any agreement between the Parties to enter into any such relationship, but not for any other purpose.

It is acknowledged that both parties operate in similar areas of the healthcare industry and that employees responsible for those operations will be some of the same employees reviewing the Disclosing Party's Confidential Information. Nothing contained herein shall be construed as preventing employees of the Receiving Party who had access to Confidential Information of the Disclosing Party from using that information retained in their unaided memory, without reference to any of the Disclosing Party's Confidential Information, as part of their general skill, knowledge, talent and expertise. It being understood that, for the avoidance of doubt, the prior sentence shall not entitle any employees of the Receiving Party to use and/or disclose any Confidential Information in breach of any of the provisions of this Agreement or in violation of the purpose herein, including information related to Chemistry, Manufacturing and Control of defibrotide.



4. **Ownership and Return of Confidential Information.** All Confidential Information of each of the Parties, as Disclosing Party, and any Derivatives thereof whether created by such Disclosing Party or the other Party, as Receiving Party, shall remain the property of the Disclosing Party, and no license or other rights to such Disclosing Party's Confidential Information or Derivatives is granted or implied hereby. For purposes of this Agreement, "Derivatives" shall mean: (a) for copyrightable or copyrighted material, any translation, abridgment, revision or other form in which an existing work may be recast, transformed or adapted; (b) for patentable or patented material, any improvement thereon; and (c) for material which is protected by trade secret, any new material derived from such existing trade secret material, including new material which may be protected under copyright, patent and/or trade secret laws. All materials (including, without limitation, documents, drawings, models, apparatus, sketches, designs, lists and all other tangible media of expression) furnished by each of the Parties, as Disclosing Party, to the other Party, as Receiving Party, shall remain the property of such Disclosing Party. At such Disclosing Party's request and no later than five (5) days after such request, such Receiving Party shall promptly destroy or deliver to such Disclosing Party, at the Receiving Party's option, (a) all materials furnished to such Receiving Party by such Disclosing Party, (b) all tangible media of expression in such Receiving Party's possession or control to the extent that such tangible media incorporate any of such Disclosing Party's Confidential Information, and (c) written certification of such Receiving Party's compliance with such Receiving Party's obligations under this section 4. Notwithstanding the foregoing, the Receiving Party shall be permitted to retain one (1) copy of the Disclosing Party's Confidential Information as necessary to ensure compliance by the Receiving Party with this Agreement and to ensure compliance with applicable law, rules or regulation in any case without prejudice to the obligation of the Receiving Party to comply with all provisions set forth herein.
5. **Required Disclosure.** The Receiving Party shall promptly notify the Disclosing Party in writing if any Confidential Information may be required to be disclosed by the Receiving Party pursuant to any provision of law, rule, regulation, judicial order, administrative order, subpoena, interrogatory, discovery request, investigative demand or other legal requirement or legal process or binding order of a competent authority. The Receiving Party will be permitted to furnish and disclose such Confidential Information required to be disclosed, provided, however, that Receiving Party shall provide reasonable cooperation with the Disclosing Party so that the Disclosing Party may seek an appropriate protective order.
6. **No License.** Nothing contained herein shall be construed to grant a Receiving Party any immunity or license under any intellectual property right of the Disclosing Party.
7. **Term of Obligation.** The Parties' obligations concerning nondisclosure of confidential information contained in the above paragraphs shall continue for ten (10) years from the Effective Date, and then terminate.



8. **No Violation.** Each Party represents that its compliance with the terms of this Agreement will not violate any duty which such Party may have to any other person or entity, including obligations concerning providing services to others, confidentiality of proprietary information and assignment of inventions, ideas, patents or copyrights.
9. **Securities Laws.** Each Party hereto agrees not to use any Confidential Information in violation of applicable securities laws. Each Party is aware, and will advise its employees and independent contractors who are informed of the matters that are subject to this Agreement, of the restrictions imposed by the United States securities law on the purchase and sale of securities by any person who has received material, non-public information from the issuer of such securities and on the communication of such information to any other person when it is reasonably foreseeable that such other person is likely to purchase or sell such securities in reliance upon such information.
10. **Injunctive Relief.** A breach by either Party of any of the promises or agreements contained herein may result in irreparable and continuing damage to the other Party for which there may be no adequate remedy at law, and such other Party shall be entitled to seek injunctive relief and/or a decree for specific performance, and such other relief as may be proper (including monetary damages if appropriate).
11. **General.**
  - a. This Agreement shall not be assigned by either Party without the written consent of the other except in connection with the transfer of substantially all of the assets or business of such Party. Subject to the foregoing, this Agreement shall inure to the benefit of and be binding upon the successors and permitted assigns of the Parties.
  - b. This Agreement shall be construed and interpreted in accordance with the laws of Switzerland, Court of Zug, without regard to its principles concerning the application of laws of other jurisdictions.
  - c. This Agreement is the entire agreement between the Parties concerning the disclosure of confidential information and supersedes all prior oral and written agreements between them. No waiver, alteration or cancellation of any of the provisions of this Agreement shall be binding unless made in writing and signed by the Party to be bound.
  - d. If any portion of the Agreement is held to be invalid or unenforceable by a court of competent jurisdiction, the remaining provisions of this Agreement shall remain in full force and effect without the invalid of unenforceable provisions.
  - e. This Agreement shall not be construed, by implication or otherwise, as an obligation by either Party to enter into any further agreement relating to the other Party's Confidential Information and/or to proceed with the Purpose. Each Party reserves the right, at its sole and absolute discretion, to suspend or terminate any discussions relating to the Purpose (and if it does so suspend or terminate such discussions prior to the execution of a definitive acquisition agreement, it shall have no liability whatsoever vis-à-vis the other Party as a result of any such suspension or termination of such discussions relating to the Purpose).



- f. To the extent that any Confidential Information includes materials or other information that may be subject to the attorney-client privilege, work product doctrine or any other applicable privilege or doctrine concerning any Confidential Information or any pending, threatened or prospective action, suit, proceeding, investigation, arbitration or dispute, it is acknowledged and agreed that the Parties have a commonality of interest with respect to such Confidential Information or action, suit, proceeding, investigation, arbitration or dispute and that it is the Parties' mutual desire, intention and understanding that the sharing of such materials and other information is not intended to, and shall not, affect the confidentiality of any of such materials or other information or waive or diminish the continued protection of any of such materials or other information under the attorney-client privilege, work product doctrine or other applicable privilege or doctrine. Accordingly, all Confidential Information that is entitled to protection under the attorney-client privilege, work product doctrine or other applicable privilege or doctrine shall remain entitled to protection thereunder and shall be entitled to protection under the joint defense doctrine, and the Parties agree to take all measures necessary to preserve, to the fullest extent possible, the applicability of all such privileges or doctrines.
- g. This Agreement may be executed in several counterparts, each of which shall constitute an original and all of which, when taken together, shall constitute one agreement. The exchange of a fully executed Agreement (in counterparts or otherwise) by electronic transmission or by facsimile shall be sufficient to bind the parties to the terms and conditions of this Agreement.
- h. Each of the Parties hereby represents and warrants that the official signing this Agreement on behalf of such Party has the power to do so on behalf of such Party.

IN WITNESS WHEREOF, the Parties have executed this Agreement effective as of the Effective Date.

**Gentium SpA**

**Jazz Pharmaceuticals plc**

By: /s/ Khalid Islam  
Name: Khalid Islam  
Title: President and Chief Executive Officer

By: /s/ Fintan Keegan  
Name: Fintan Keegan  
Title: EVP Technical Ops